The Trenches: Interesting Cases in a University Setting

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Financial Disclosures

- none

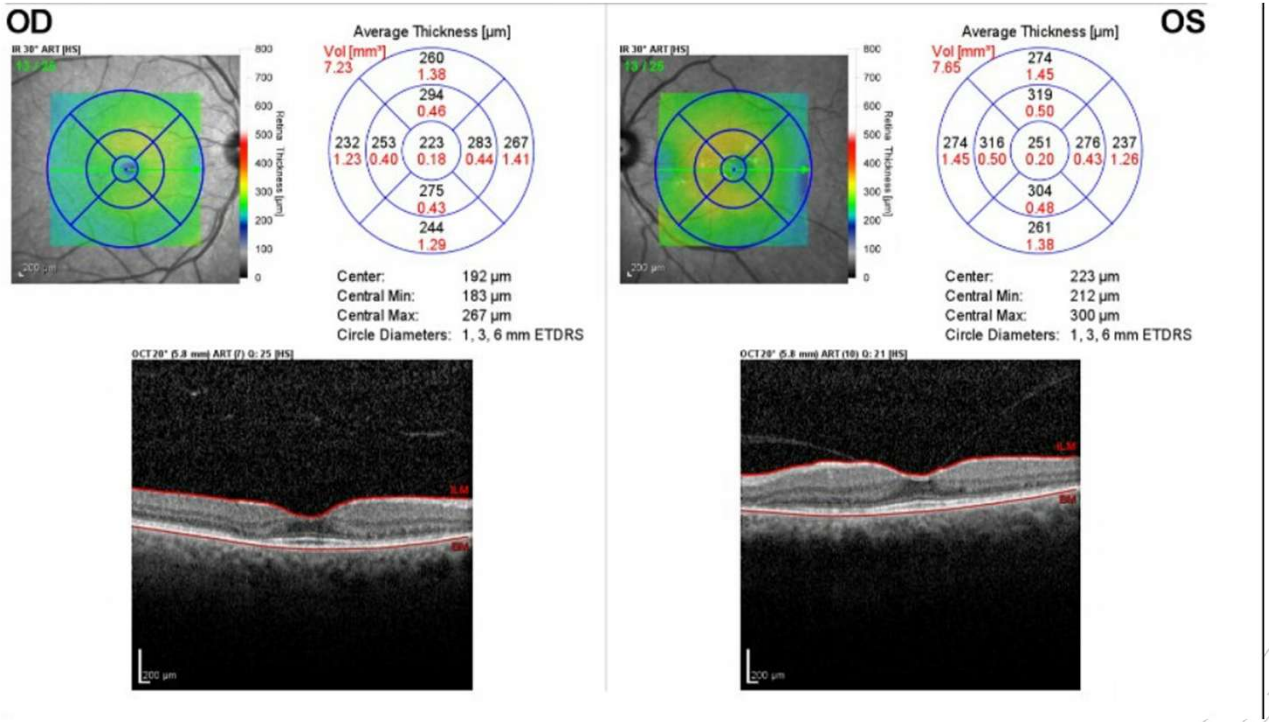
University of Kentucky

- Over 30 providers
  - Main campus – Shriner's Hospital in Lexington, KY
  - Optometry and ophthalmology
  - Multiple subspecialties
  - Many community clinics and satellites
- Education meets patient care
  - Optometry students
  - Optometry resident
  - Ophthalmology residents
  - Medical students
  - Undergraduate and high school students

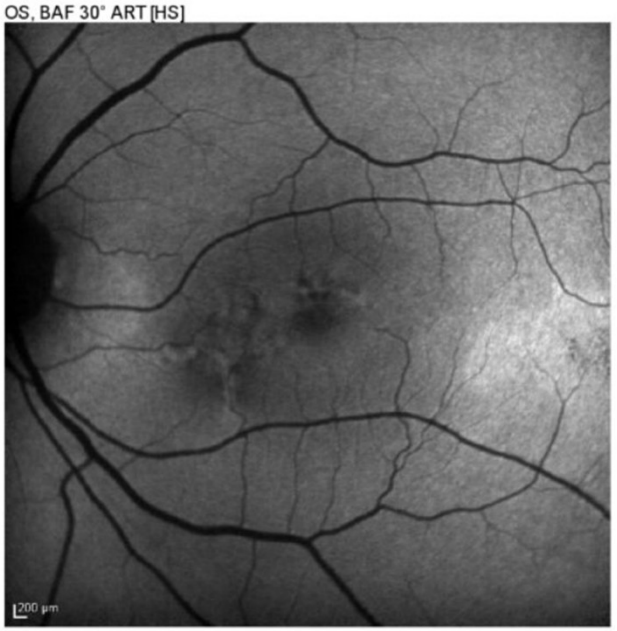
Hydroxychloro-collaboration

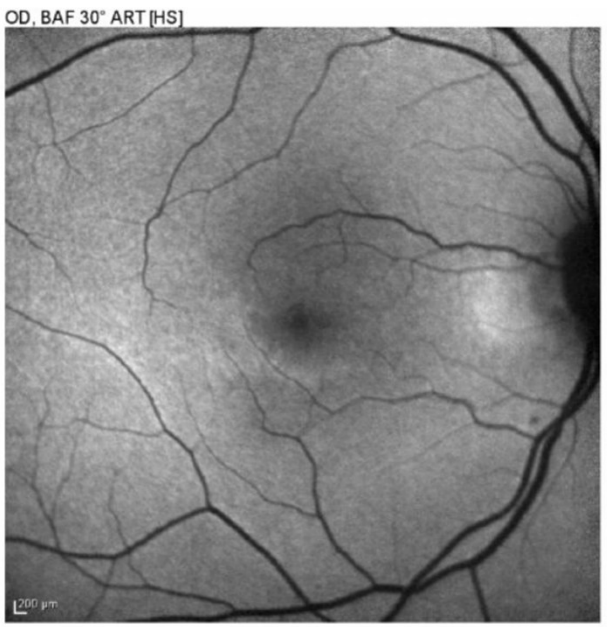
- 71 yo M presents for DM eye exam
- Hx psoriatic arthritis since 1996
- Reported hx HCQ toxicity in 2017 – has since d/c
  - Also hx CSCR OS
- VA 20/25 OD, 20/25+2 OS
- PERRL, -APD OU, CVF full OU, EOMs full OU
- IOP 16/14 tonopen
- SLE remarkable for MGD, combined cataracts OU
- Fundus exam remarkable for pigment mottling and blunted FLR OU

Macular OCT


OD
Average Thickness [µm]
Vol [mm³]
7.23
260
1.38
294
0.46
232
1.23
253
0.40
223
0.18
283
0.44
267
1.41
275
0.43
244
1.29
Center: 192 µm
Central Min: 183 µm
Central Max: 267 µm
Circle Diameters: 1, 3, 6 mm ETDRS
OS
Average Thickness [µm]
Vol [mm³]
7.65
274
1.45
319
0.50
274
1.45
316
0.50
251
0.20
276
0.43
237
1.26
304
0.48
261
1.38
Center: 223 µm
Central Min: 212 µm
Central Max: 300 µm
Circle Diameters: 1, 3, 6 mm ETDRS

Autofluorescence





Back in 2017…

- Initially prescribed HCQ for his PA in 1996, which worked for a long time
- Failed two visual field screenings for HCQ toxicity, referred to University of Michigan
- Stopped HCQ per OD instructions about 6-8 weeks prior to exam, reporting significantly increased joint pain
- Subjectively reporting increased blurred vision over the last couple years
- "200 mg BID for total daily dose of 400 mg. Daily dose equivalent to 4.2mg/kg. **Cumulative dose - enormous** (20years x 365days/year x 400 mg/day =29,200 g"
- Stay off HCQ and monitor

Back to 2021

- Thankfully stable since Michigan days
- Will need to watch closely given hx CSCR OS confounding retinal imaging
- Communicated with rheumatology at Lexington Clinic (and also UofM)
- Currently well-managed with Cosentyx
- Monitor with yearly DFE, mac OCT, OCT FAF

Plaquenil Research Update

- Classically considered an outer retinal disease, but now shown to affect inner retinal layers as well
- May be stored in fat in addition to melanotic tissue, liver, and kidneys
- Earlier detection
  - Fundus autofluorescence
  - OCT-A
  - Genetic markers

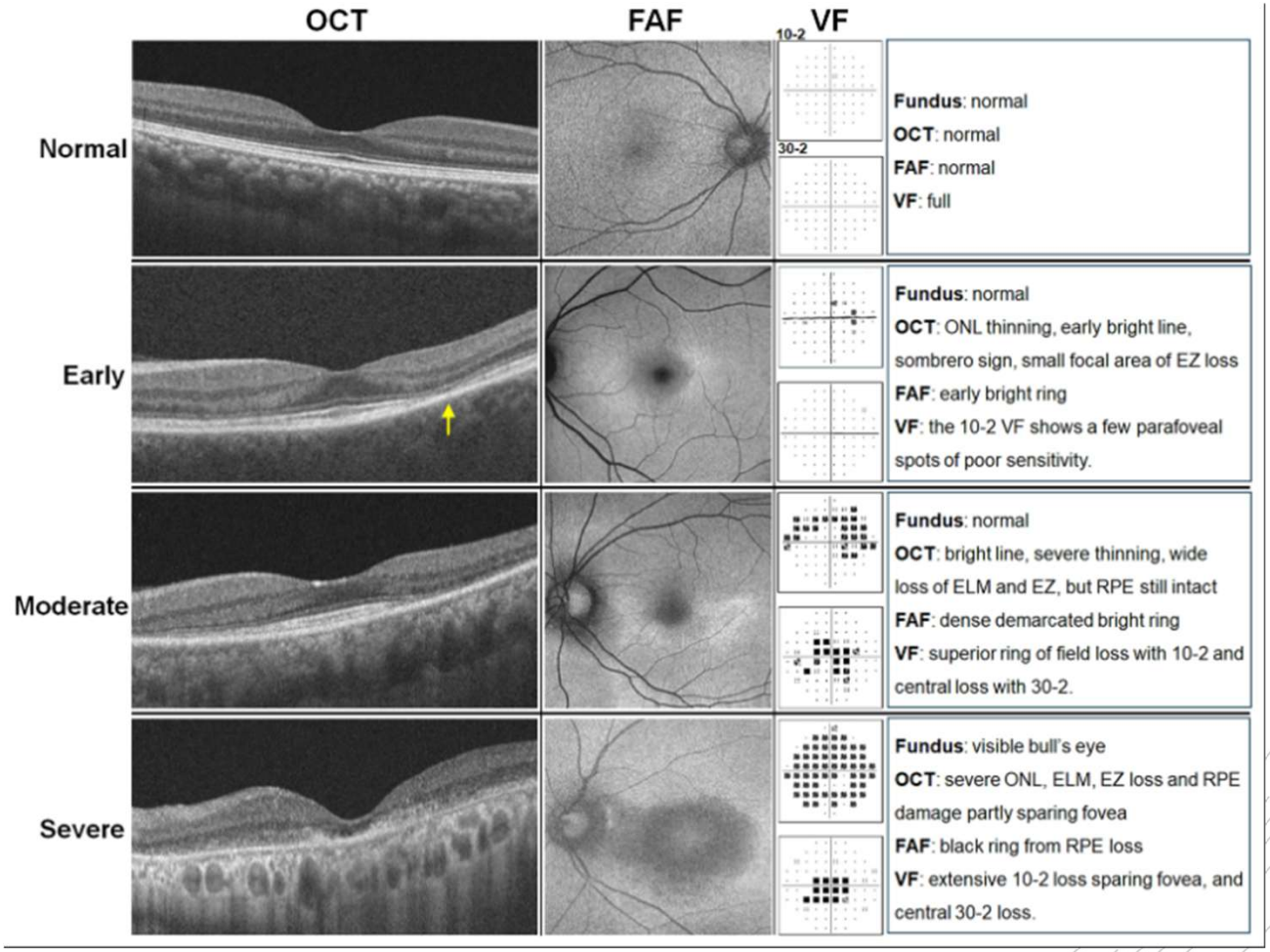
Special AAO Report: Recommendations on Screening for Hydroxychloroquine Retinopathy (2025 Revision)

- Recommended daily dosage remains unchanged at ≤5.0 mg/day/kg real weight
  - Keep under 400mg/day for severely obese patients
- Mild retinopathy changes may not progress if HCQ is stopped with only early OCT changes, but may still progress for many years if severe
- Additional risk factors
  - Renal disease
  - Tamoxifen use
  - Initiation of HCQ at older ages
  - Any macular disease

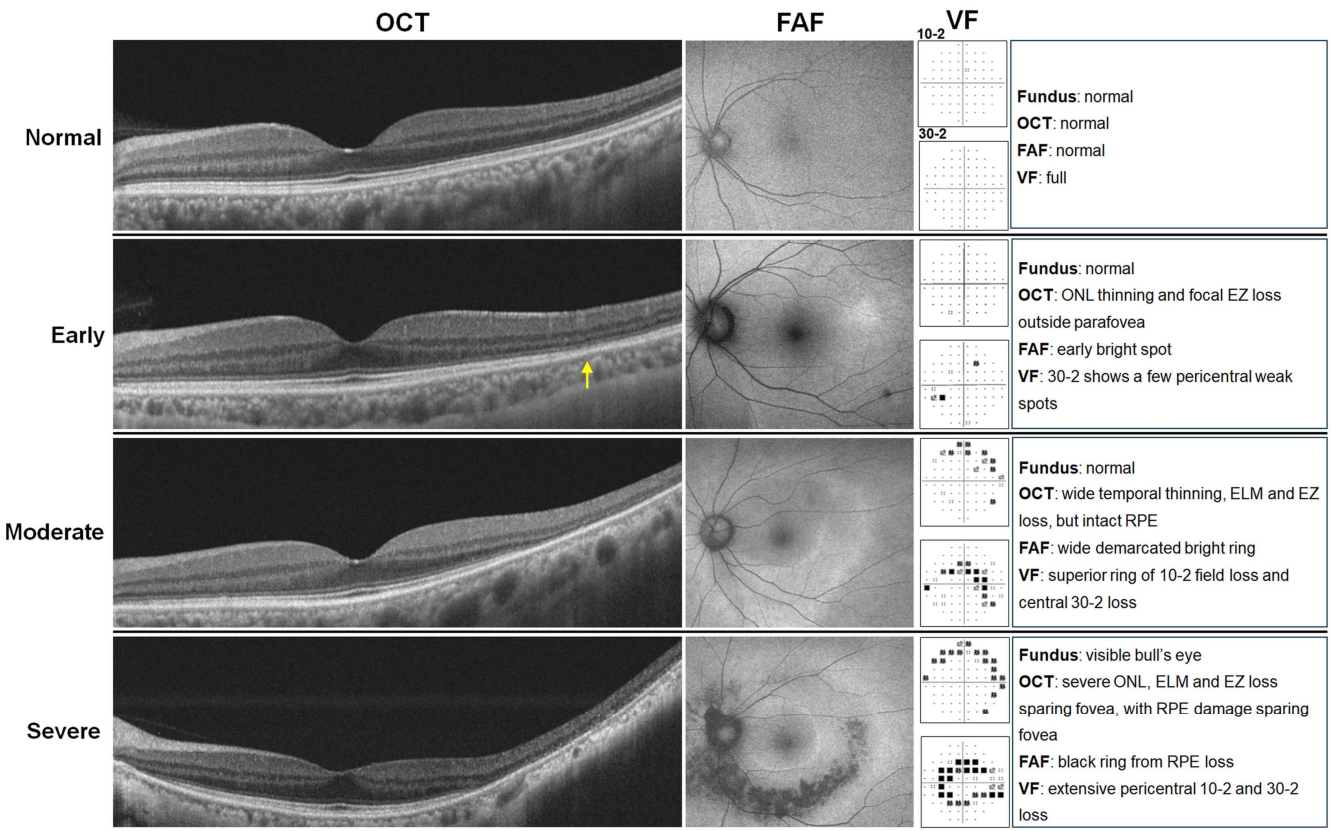
Special AAO Report: Recommendations on Screening for Hydroxychloroquine Retinopathy (2025 Revision)

- Baseline examination recommended shortly after initiating treatment
  - OCT, wide pattern fundus autofluorescence
  - 10-2 visual field now "secondary"
    - Also mfERG
    - Consider 24-2C
  - Recommend annual screening for first five years, but may be deferred if no other risk factors present
- Different patterns of fundus changes
  - European, Middle Eastern – parafoveal distribution
  - East Asian – extramacular, pericentral
  - Black Americans – greater tendency toward pericentral disease than white Americans

Plaquenil Toxicity - Parafoveal



Plaquenil Toxicity - Pericentral



When To Stop

- When changes are found, conversations should be had
- Collaboration with patient and rheumatologist/prescribing physician
- May not have good alternatives
  - Lupus, chronic pain
- Will get harder as earlier detection becomes possible

Back To the Future

- Pt presented again in December 2025 for annual DM exam
- VA 20/40 OD, 20/20- OS
  - Consistent with worsening cataracts
- Exam otherwise stable, but new drusen present OS
- Still controlled with Cosentyx and indomethacin
  - Still follows with University of Michigan rheumatology
- Now also managing DM with Ozempic in addition to metformin
- Always watching…

Always Ask the Wife

- 62 yo M complains of OD swelling x 4 days
- Hx MGD and preseptal cellulitis
- Complains of pressure sensation, clear discharge throughout the day
- Previous episodes treated with amoxicillin PO and erythromycin ophthalmic ointment
- Adherent to warm compresses
- "My wife told me to tell you that I have chronic sinusitis."
  - Nasal polyps, sinus surgery 10 years ago

Clinical Findings

- VA 20/20 OD, OS
- PERRL, -APD OU; CVF full OU; EOMs full OU
- IOP 16/14 tonopen
- SLE remarkable for reactive edema RUL, MGD, 1+ papilla OD>OS
- OD does appear to be slightly proptotic when speaking with pt
  - Exophthalmometry – 19mm OD, 15mm OS, base 99
- Undilated posterior pole exam unremarkable/stable

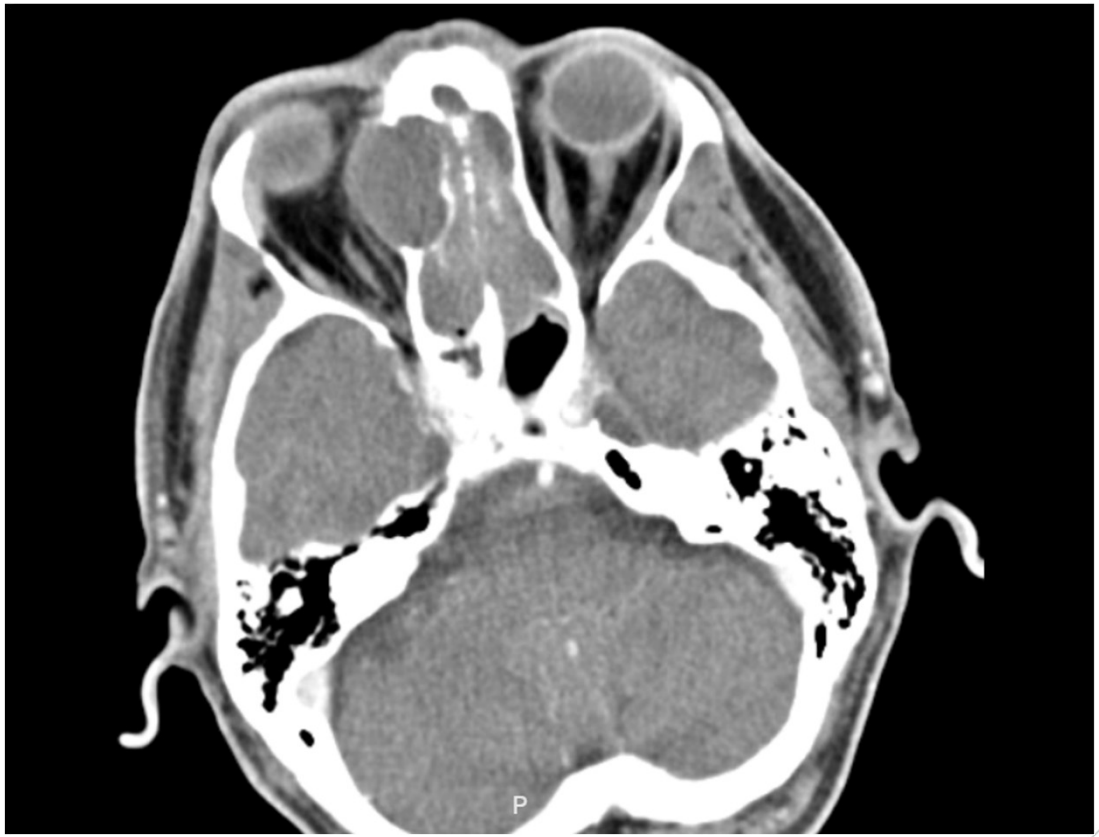
Assessment and Plan

- Given proptosis and reported sinus history, ordered CT orbits w/ contrast
- Rx maxitrol ophthalmic ung BID OD and cephalexin 500mg BID PO x 7 days
- Continue warm compresses, gentle lid hygiene, artificial tears
- Monitor x 2 weeks

Follow-Up

- Presented 2 weeks after with much improved surface, but still proptotic
- CT orbits performed the day prior and had not had report finalized, but images available

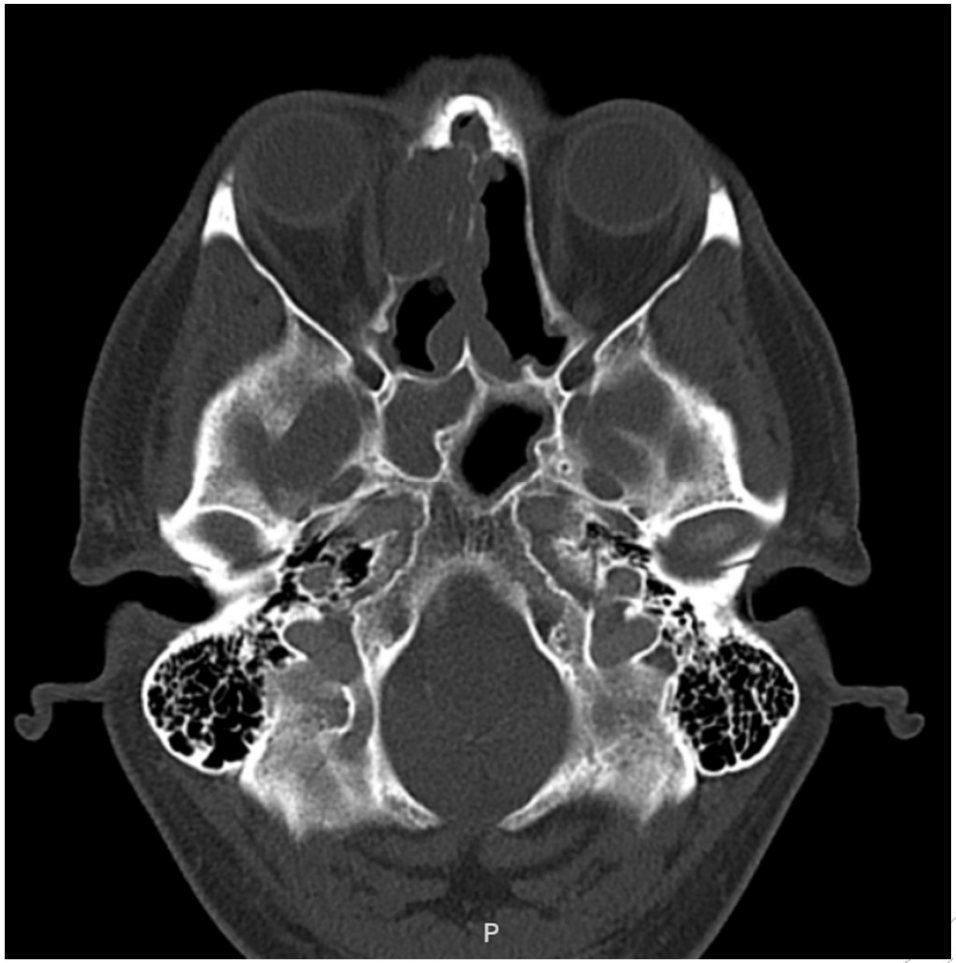
CT orbits w/ IV Contrast



Plan

- Primary differential includes mucocele, but infection or malignancy not ruled out
- Recommend MRI to differentiate tissue
  - Offered outpatient or ED, pt chose ED
- Consulted ENT and oculoplastics aware
- MRI more clearly defines as mucocele, ENT prescribed prednisone and clindamycin
- Inpatient repeat CT shows improvement but still present and proptotic

Repeat CT



Outpatient ENT Consultation

- Improved with medical treatment, no visual/ophthalmologic symptoms now except for proptosis
- In-office nasal endoscopy performed in which forceps were used to open mucocele and evacuate mucopurulence
- Continue nasal care and prednisone
  - Had finished oral abx
- ENT follow-up and subsequent optometry visits show resolution of proptosis and any visual complaints, though sinus disease persists

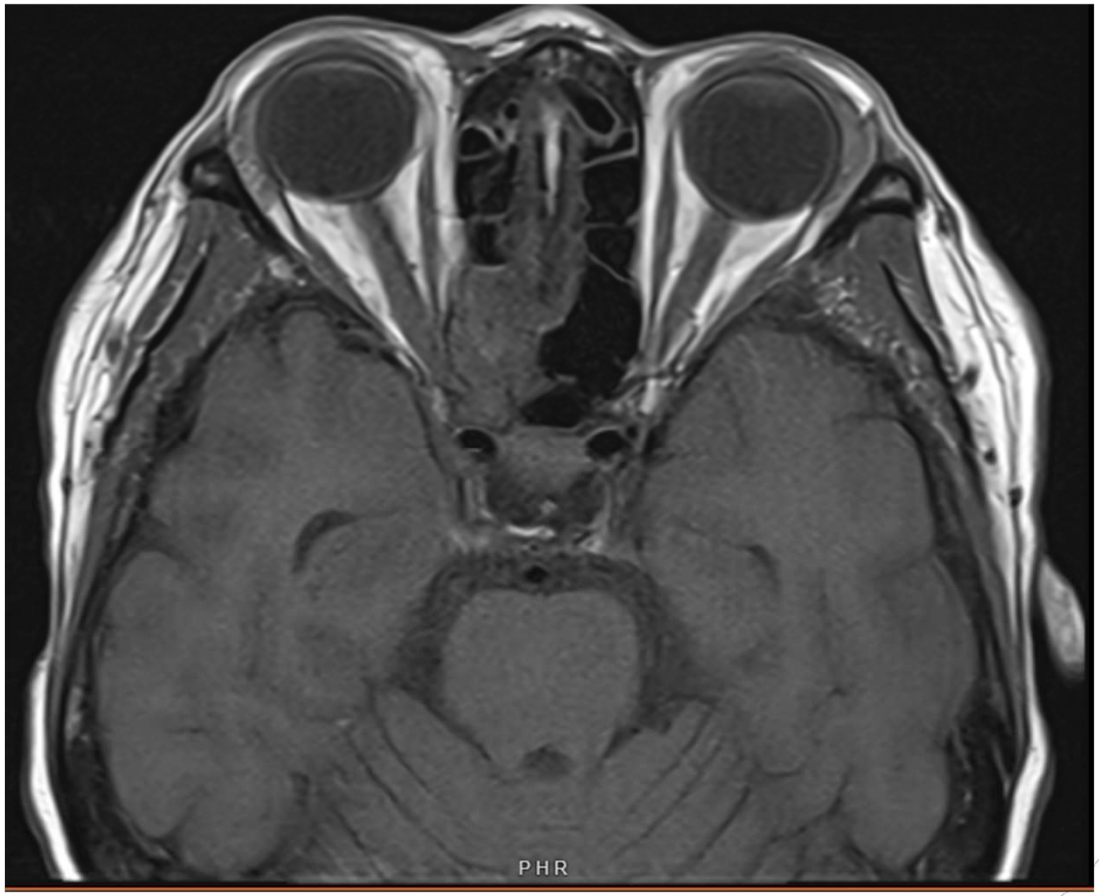
What is a Mucocele?

- Benign, mucus-containing cysts formed due to obliteration of sinus ostium
- Most commonly found in frontal sinus
- Capable of intraorbital and intracranial spread
  - Meningitis, brain abscess, loss of vision
  - Bone resorption and new bone formation
- Paranasal sinuses often treated endoscopically
- There have been cases where an orbital decompression was needed

One Battle After Another

- 69 yo F presents for follow-up from hospital admission for optic neuritis (11/2025)
- Hx polycythemia vera with secondary myelofibrosis
- Had received allogenic stem cell transplant 09/2025
  - Neutropenic fever with no clear cause, treated with IV abx
  - Right eye swelling and headache, CT orbits revealed sinusitis, treated conservatively as signs and symptoms improved
  - At follow-up with heme-onc in 10/25, pt complained of new diplopia, blurred vision, and right-sided headache
  - Admitted with MRI orbits which revealed invasive fungal sinusitis with bony dehiscence and enhancement of right optic nerve

MRI Orbits



Inpatient Consultation – 10/2025

- VA 20/50 ph 20/20-2 OD, 20/30 ph 20/20-2 OS
- +APD OD, CVF full
- Pain with lateral gazes OD
- Color 6/11 OD, 11/11 OS, +red desaturation
- SLE unremarkable, fundus exam remarkable for several small yellow lesions surrounding optic nerve
- Dx with optic perineuritis, MRI brain and MRV ordered due to possible APMPEE; consulted Infectious Disease and Neurology

Inpatient Course

- 10/11- ENT performed nasal endoscopy without evidence of IFS
- 10/12 with acute worsening of vision
  - Repeat MRI showed dural enhancement and opacified right posterior ethmoid with signal changes worrisome for fungal colonization
  - LP w/ CSF studies performed
- 10/13 with mild improvement in vision and symptoms - suspect due to dose of steroids given the evening prior
- 10/14 stable exam
- 10/15 stable exam, but subjective worsening in vision with worsening "greying" in central vision
- 10/17 with stable exam, but again subjective worsening of central scotoma.
  - MRI orbits showing worsening optic nerve enhancement suggestive of optic neuritis, right posterior ethmoid sinus suspicious for invasive fungal sinusitis
- 10/17 - s/p Right FESS with maxillary antrostomy and total ethmoidectomy with sphenoidotomy with tissue removal. Intraoperative findings consistent with fungal ball and superimposed bacterial infection
  - No evidence of invasive fungal sinusitis
- 10/20 : now s/p surgery with ENT, with improvement in vision and other symptoms

Back to 11/25

- Doing well following discharge
- VA 20/20 OD, OS
- Color plates normal OD, OS
- APD not visualized
- Exam unremarkable
- Follow-up 6-8 weeks

Interim

- Hospitalized 12/2025 due to rising liver function tests, concern for hepatic graft versus host disease – had to cancel scheduled follow-up
  - Had completed post-transplant cyclophosphamide, abatacept, and tacrolimus
  - On infection prophylaxis
- Saw neuro-ophthalmology in interim
  - "Extremely complex case that is followed by ENT, Infectious Disease, Optometry and Oncology"
  - Resolution of optic perineuritis, likely secondary to fungal sinusitis and not with any primary etiology (eg demyelination)
  - Significant dryness symptoms – initiated Xiidra

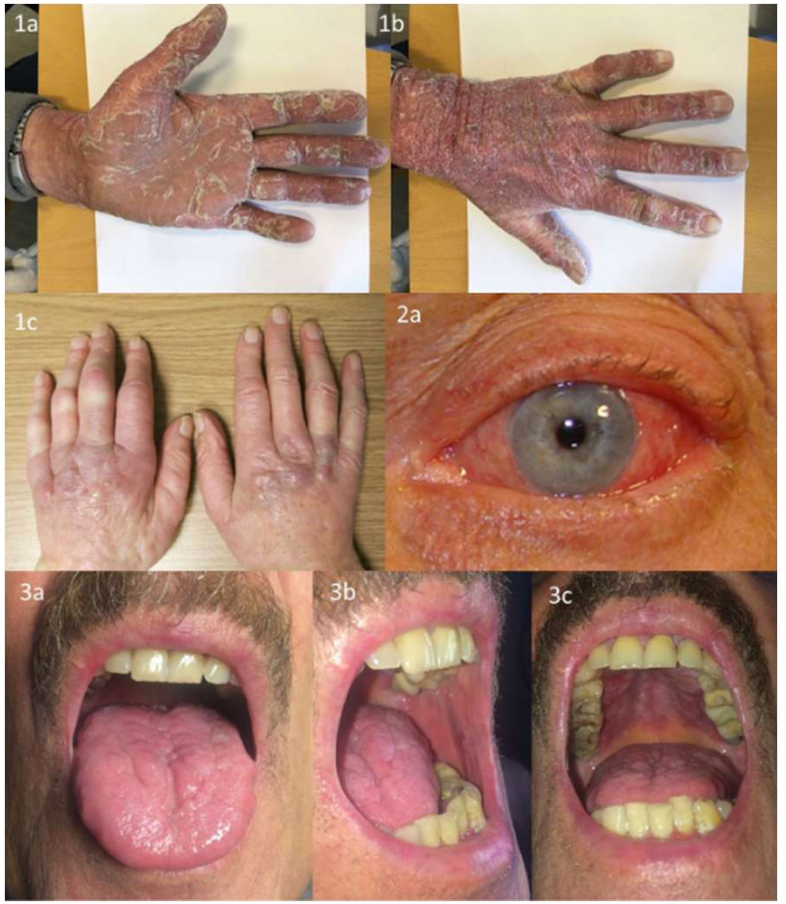
Optometry Visit 03/2026

- Reports worsening pain around OD radiating to lower back, blurred vision OD
- Has noted improvement since starting Xiidra
- VA 20/20-2 OD, 20/20 OS
- Preliminary testing unremarkable
- SLE significant for 3+ diffuse PEE, tbut <5sec OD, 1+ diffuse PEE, tbut <5 sec OS
- Severe dry eye vs ocular GVHD
  - Continue Xiidra, add Miebo QID and erythromycin ophthalmic ung nightly

Graft Versus Host Disease

- Transplanted donor immune system recognizes healthy recipient cells as foreign, leading to inflammation and tissue destruction
- Can lead to severely reduced quality of life, blindness, and reduced sleep quality
- Allogenic HSCT uses donor immunity to eradicate disease, which can lead to GVHD
- 30-70% of patients who receive HSCT will develop systemic chronic GVHD
- Primarily affects liver, skin, mouth, and eyes
- Pathophysiology not entirely understood
- Fibroblasts found in lacrimal glands, CD4+ and CD8+ cells found in periductal areas
- Almost always begins within 1 year of transplant

GVHD



Ocular GVHD

- AcuteGVHD
  - Conjunctival inflammation
  - Corneal epithelial sloughing
  - Pseudomembranes
- ChronicGVHD
  - Keratoconjunctivitis sicca – most common
  - Meibomian gland dysfunction
  - Secondary corneal infections/ulcers
    - Can lead to perforation in worst cases
  - Cicatricial conjunctivitis
    - Entropion/ectropion, trichiasis

Scoring Criteria

	SCORE 0	SCORE 1	SCORE 2	SCORE 3
EYES *Keratoconjunctivitis sicca (KCS) confirmed by ophthalmologist:* ☐ **Yes** ☐ **No** ☐ **Not examined**	☐ No symptoms	☐ Mild dry eye symptoms not affecting ADL (requirement of lubricant eye drops ≤ 3 x per day)	☐ Moderate dry eye symptoms partially affecting ADL (requiring lubricant eye drops > 3 x per day or punctal plugs), **WITHOUT** new vision impairment due to KCS	☐ Severe dry eye symptoms significantly affecting ADL (special eyeware to relieve pain) **OR** unable to work because of ocular symptoms **OR** loss of vision due to KCS

☐ *Abnormality present but explained entirely by non-GVHD documented cause (specify):*

Treatment Options

- Artificial tears
- Punctal occlusion
- Corticosteroids
- Immunomodulating drops
  - Cyclosporine, tacrolimus
- Oral antibiotics for MGD
- Serum tears
- Contact lenses
  - Soft or scleral lenses
  - Filamentary keratitis
  - Recurrent trichiasis
- Tarsarraphy
- Amniotic membrane

Patient Update – 04/2026

- Still symptomatic, but overall improved
- VA 20/20 OD, OS
- Preliminary testing unremarkable
- Still has diffuse PEE OD>OS with reduced tbut
- Consider serum tears, but due to anemia will want to hold off for now
- Punctal plug inserted RLL only

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Thank you!