

AVOID!

MISTAKES NOT TO MAKE IN RETINAL AND ANTERIOR SEGMENT DISEASE

Joseph Sowka, OD

OOPS!

MISTAKES HAPPEN.

MISTAKES NOT TO MAKE

- Undertreating anterior uveitis
- Not dilating eyes with anterior uveitis
- Not considering all possible causes of infectious keratitis
- Not considering age-appropriate causes of retinal vein occlusions
- Not referring TIA and RAO immediately to a stroke unit

MISTAKE NOT TO MAKE

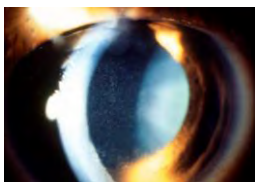
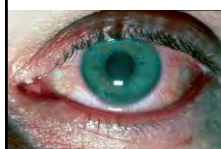
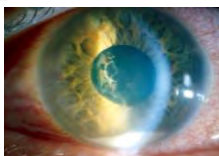
- Undertreating anterior uveitis

ANTERIOR UVEITIS

- **Inflammation of the anterior uveal structures**
 - Increased vascular permeability
 - Breakdown of blood- aqueous barrier
- **Primary inflammation in absence of precipitating conditions**
 - Human leukocyte antigen B27 (HLA-B27) is a protein located on the surface of white blood cells
 - HLA-B27 contributes to immune system dysfunction. The presence of HLA-B27 can cause immune system to attack healthy cells. This results in autoimmune disease or immune-mediated disease, such as juvenile rheumatoid arthritis or ankylosing spondylitis.

ANTERIOR UVEITIS

- Pain, photophobia, lacrimation
- Circumlimbal flush
- Cells & flare in anterior chamber
- Possible hypopyon
- Posterior and anterior synechia
- IOP typically decreased, but may be elevated
- Keratic precipitates



ANTERIOR UVEITIS: MANAGEMENT

- **Remove antigen**
 - Trauma, corneal disease, infection
 - Antigen not identified in autoimmune, idiopathic, recurrent uveitis. Difficult to manage. Must beat down inflammation
- **Strong cycloplegics**
- **Immunosuppressants**
 - Corticosteroids
 - Systemic immunosuppressants
 - Chemotherapeutic agents



MANAGEMENT CONSIDERATIONS:

- **Rapid control of inflammation in acute anterior uveitis is mandatory**
- **Must break attack early, or longer duration of therapy will be needed**
 - Steroids Q1H while awake initially, then decrease to Q2H
 - May need higher initial dose
- **Taper steroids only when there are ZERO cells present**

MANAGEMENT CONSIDERATIONS:

- Each acute episode should only last 6-8 weeks.
- Longer indicates that initial therapy not aggressive enough
- Painful with lots of inflammation – recheck with 72 hours

MANAGEMENT CONSIDERATIONS:

- **Allowing chronic inflammation causes damage and vision loss**
 - Under treated
- **Limping along with 1+ cells not acceptable**
 - However, may be necessary
- **Goal of treatment**
 - No inflammation
 - No steroids
 - No complications
- **Chemotherapy may be necessary**

CLINICAL PEARLS

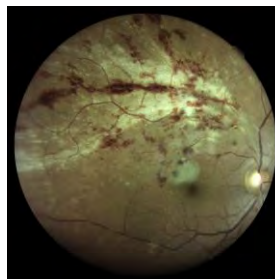
- Initial aggressive anti-inflammatory therapy is key to success.
- Follow closely...but not too closely.
 - Daily f/u of uveitis is like watching paint dry
- **Never taper steroids as cells decrease- taper only when no cells are present. Otherwise there will be a rebound and prolonged course**

MISTAKE NOT TO MAKE

- **Not dilating anterior uveitis**

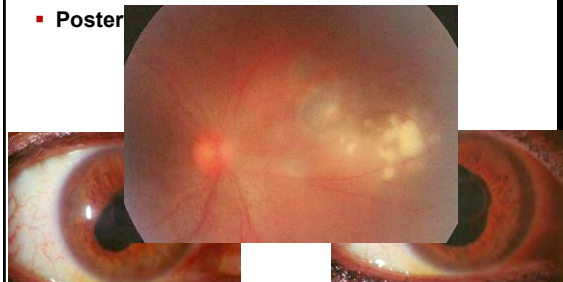
46 YOM

- Anterior uveitis OD with mild reaction



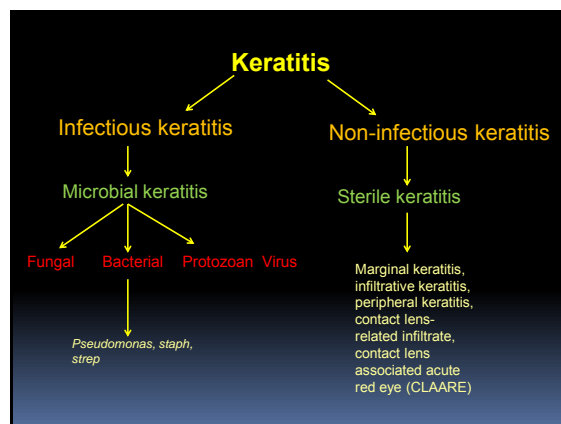
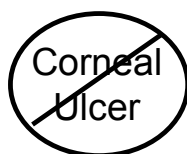
40 YOF

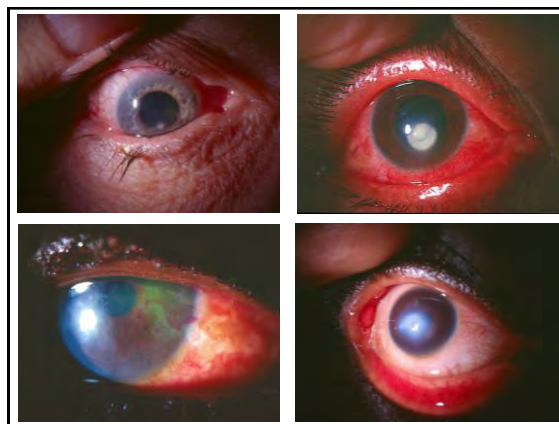
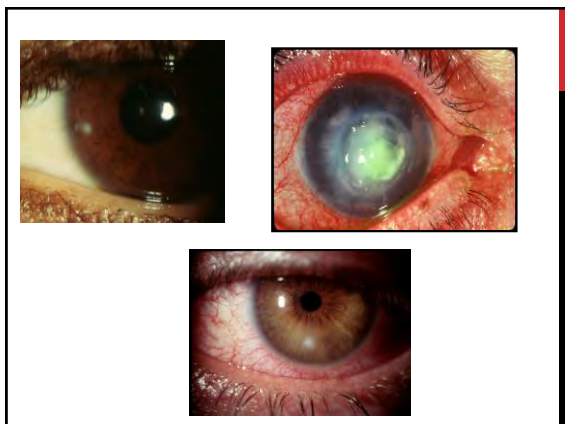
- Anterior uveitis OS
- IOP 03 mm
- Posterior

**MISTAKE NOT TO MAKE**

- Not considering all possible causes of infectious keratitis

- 19 year old female
- CC: developed a red, painful right eye while wearing contact lenses
 - Goes to the emergency room where they patch her eye with gentamicin after trying to remove "white foreign body".
- Med Hx: (-); No meds; NKDA
- VA: PH 20/100 OD, 20/20 OS





ALIEN KERATOPATHY?



ACANTHAMOEBA:

Opportunistic free-living soil amoebae

- Extremely hardy; can withstand extremes in temperature and survive numerous environments

Presentation is variable

- Typically unilateral, painful red eye
- Tearing, reduced vision, photophobia

Ring infiltrate...?

Atypical corneal lesions; mimics HSV

- Non-specific PEK or pseudodendrite
- Irregular epithelial defect
- Disciform stromal keratitis

Radial perineuritis (~1/3rd of cases)

Microcystic or bullous keratopathy

Anterior uveitis +/- hypopyon



GETS BETTER WITH STEROIDS!

- 30 year old White male
- Painful, red left eye x 2 weeks
- Treated previously for "corneal abrasion"
 - Gentamicin gtt and ung with patching QHS
 - Minimal epitheliopathy
- Treated subsequently with Voltaren, debridement, bandage lens, TobraDex, erythromycin ointment
 - Enjoyed TobraDex



HERPES SIMPLEX DISCIFORM KERATITIS: SYMPTOMS & SIGNS

Pain	Discrete disc shaped areas of focal stromal edema
Photophobia	Stromal infiltration
Lacrimation	Central or peripheral
Vision loss	Epithelium intact
	Avascular
	May be severe with corneal melting



GETS WORSE WITH STEROIDS

- 22 YOM- Contact lens wear
- Feels he scratched eye trying to remove lens on canoe trip
- Seeks care from ED
 - Treated with ciprofloxacin ab drops and ointment
- Getting worse x 2 weeks
- Doctor adds corticosteroid- Worsens immediately



FUNGAL KERATITIS

- ocular trauma--particularly if organic vegetative matter is involved
- topical steroid therapy
- ocular or systemic immunosuppressive diseases.
- agricultural and tropical environments.
- altered epithelial barrier increases the threat.
- filamentary fungal infections initially produce a feathery, branching pattern.
- The cornea takes on a dull gray appearance with possible heaping of epithelium and a dry, rough texture.
- Typically a severe anterior uveitis and plasmoid aqueous with hypopyon appears.

MISTAKE NOT TO MAKE

- Not considering age-appropriate causes of retinal vein occlusions

THE CASE OF THE COLORED FLASHING LIGHTS

- 45 YOHF presented with colored "map-like" phosphenes and small black flashing spots OD x two weeks
- Noted that she had to "look between the lights" to see out of her right eye.
 - 6/6 OD, OS
- Medical history was unremarkable except for treated migraines
- Lost 1 pregnancy



CASE CONTINUED

- She returned four days later complaining of decreased vision in the right eye, which had reduced to counting fingers at ten feet.
 - Macular edema, more extensive hemorrhaging, cotton wool spots, disc edema and dilated vessels
- Underwent IV Kenalog injections and showed improved vision of 6/24 OD during follow up examinations.
 - Released by retinal specialist
 - No medical evaluation

Now
What?



Are there any tests that you would like to order?

CRVO: SYSTEMIC CONSIDERATIONS

Hypertension	Diabetes
Hyperviscosity	Syphilis
CV disease	Cardiovascular disease
Sickle	Leukemia
Polycythemia	Carotid artery disease
Hyperlipidemia	Sarcoid
Autoimmune factors	Clotting abnormalities
Homocysteine	

TREATMENT & MANAGEMENT

- Referred blood work through PCP
 - DM, HTN, hypercoag, ANA, antiphospholipid antibodies, anticardiolipin, PT, PTT, ESR, CBC with diff
- Elevated erythrocyte sedimentation rate
- Mildly elevated cholesterol level.
- Elevated anti-cardiolipin IgM antibodies
 - Suggestive of antiphospholipid antibody syndrome
 - She was recommended for long term anti-coagulant therapy to prevent future thrombotic events, but patient never followed through.

CASE CONTINUED

- Seven months later the patient returned with the same signs and symptoms in her right eye.
- At this time, the vision was markedly more decreased with more evidence of ischemia
 - CF @ 6'
- She was referred to a hematologist
- Now on anti-coagulation therapy

PRIMARY ANTIPHOSPHOLIPID ANTIBODY SYNDROME

- **Thrombotic disorder**
- **Secondary antiphospholipid syndrome**
 - Associated several autoimmune diseases but most often systemic lupus erythematosus
- **Primary antiphospholipid syndrome is not associated with further systemic disease**
- **Recurrent vascular thrombosis, pregnancy loss and positive anticardiolipin or lupus anticoagulant are all characteristics of this disorder**

PRIMARY ANTIPHOSPHOLIPID ANTIBODY SYNDROME

- **The clinical criteria**
 - One or more vascular thrombotic episodes of venous, arterial or small vessel thrombosis in any organ or tissue or spontaneous abortion.
- **Laboratory testing must show persistently elevated anticardiolipin antibodies, IgG and/or IgM or lupus anticoagulant (inhibits the conversion of prothrombin to thrombin) at least six weeks apart**

PRIMARY ANTIPHOSPHOLIPID ANTIBODY SYNDROME

- **Phospholipids are identified by the body as "foreign."**
 - The antiphospholipid antibodies are produced against the "foreign" antigen.
- **The antibodies appear to react with the cell membranes causing irritation or stimulation, thus disrupting the coagulation cascade**
- **This disruption leads to abnormal blood clotting and inhibits normal phospholipid binding.**

PRIMARY ANTIPHOSPHOLIPID ANTIBODY SYNDROME

- **This abnormal or inhibition of proper phospholipid binding leads to a hypercoagulable state thus causing thrombosis.**
- **Propensity of clot formation is within the venous and arterial portions of the vascular tree, especially targeting the retinal vessels and placenta**

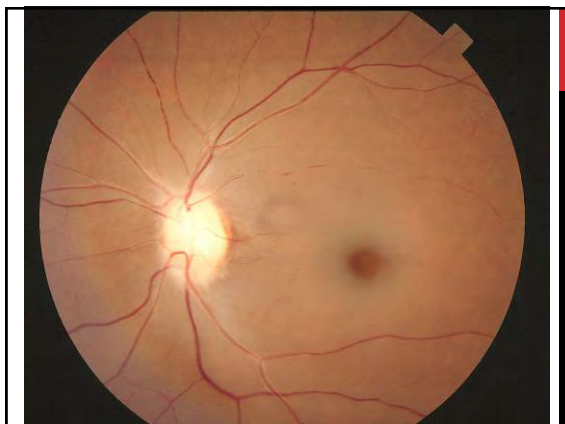
MISTAKE NOT TO MAKE

- **Not referring TIA and RAO immediately to a stroke unit**

CASE: I (DON'T) FEEL GOOD!

- **66 year old Black male**
- **CC: sudden, painless blurring OS x 3 days**
- **No previous eye or medical care**
- **Wants glasses to clear vision**
- **BVA OD 20/30, OS HM**
- **Pupils: ERRL (+) RAPD OS**
- **Good appetite, poor diet**



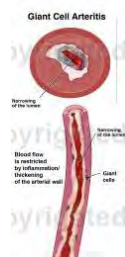


CENTRAL RETINAL ARTERY OCCLUSION (CRAO)

- Painless, sudden loss of vision
 - < 20/400 in most cases
- Retinal edema and white fundus – hypoperfusion
 - Cherry red spot
- 60's and above
- Early and late appearances
 - Initially normal fundus
 - Optic atrophy with attenuated vessels

CRAO: ETIOLOGY

- Emboli from heart or carotid lodging at lamina
- Intraluminal thrombosis
- Dissecting aneurysm
- Vasospasm
- Arteriolar necrosis
- **GIANT CELL ARTERITIS!**



CRAO: TREATMENT ?

- Paracentesis
- Carbogen
- Digital massage
- Hyperventilation
- Urokinase/streptokinase
- 1-24 hr window of opportunity
- Does anything work?

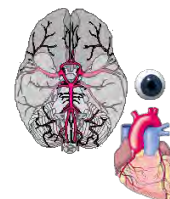


CRAO: SYSTEMIC CONSIDERATIONS

- | | |
|-------------------------------|--------------------------|
| Atherosclerosis | Hypertension |
| Carotid artery disease | Diabetes |
| GCA | Cardiac valve disease |
| Antiphospholipids ABS | Cardiovascular disease |
| Infectious endocarditis | Hyperlipidemia |
| Vasospastic disease | Disc drusen |
| Cardiac arrhythmia | Mural thrombosis |
| Clotting factor abnormalities | Hyperviscosity syndromes |

CRAO: COMPLICATIONS

- CVA
- MI
 - Main cause of death
 - 9 yr mortality 56%
- NVG
- OIS
- Fellow eye involvement if GCA cause



CRAO: MANAGEMENT

- STAT ESR and CRP for GCA
- Cardiology/ internal medicine referral
- Monitor in 3-6 mos for neovascularization
 - How common is neo?

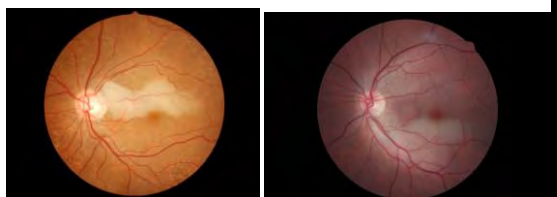


JAMES' OUTCOME

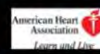
- Referred for medical care
- Diagnosed with hypertension, NIDDM, hypercholesterolemia
- Returns for ocular follow up 3 months later
 - “I’m scared”
- Several toes amputated from diabetes
- Passed away from MI within year

BRAO; CILIORETINAL AO

- BRAO nearly always embolic
- Greater risk of cardiac mortality
- Cilioretinal AO- branch of PCA- high risk of GCA



Guidelines



- Any patient with suspected TIA or those with acute retinal ischemia should be evaluated urgently in order to identify those at high risk of immediate cerebral infarction and cardiac ischemia

Guidelines for the prevention of stroke in patients with stroke or transient ischemic attack: a guideline for healthcare professionals from the American Heart Association/American Stroke Association. Stroke 2011; 42: 517-524

Adapted from Drs. Nancy Newman and Blouise; 2015

All Patients with Acute Retinal Arterial Ischemia



- MUST have immediate brain imaging
 - Brain MRI with DWI >>> Head CT
- Including patients with transient visual loss (presumed of vascular origin)



Presence of cerebral ischemia portends higher risk of stroke

Adapted from Drs. Nancy Newman and Blouise; 2015

Concurrent Acute Brain Infarcts in Patients with Monocular Visual Loss

- ¼ with acute retinal ischemia had acute brain infarction (anywhere) on brain DWI-MRI
 - Infarctions often small, multiple, ipsilateral to retinal ischemia, asymptomatic
- DWI-MRI abnormal in:
 - 33% with CRAO/BRAO vs 18% with TVL
 - 28 % with embolic vs 8% non-embolic retinal ischemia



Adapted from Drs. Nancy Newman and Blouise; 2015

Study #2

Co-occurrence of Acute Retinal Artery Occlusion and Acute Ischemic Stroke: Diffusion-Weighted Magnetic Resonance Imaging Study

JUNWON LEE*, SEUNG WOO KIM*, SUNG CHUL LEE, OH WOONG KWON, YOUNG DAE KIM, AND SUK HO RYEOH

Am J Ophthalmol 2014; 157: 1231-1238

Adapted from Drs. Nancy Newman and Biousse; 2015

Co-occurrence of acute retinal artery occlusion and acute ischemic stroke: Diffusion-weighted magnetic resonance imaging study

- 33 patients with CRAO (18) and BRAO (15)
- Evaluated similarly to acute stroke patients (DWI)
- ¼ with acute retinal ischemia had acute brain infarction (anywhere) on brain DWI-MRI
 - 5/18 CRAO; 3/15 BRAO
 - Infarctions often small, multiple, ipsilateral to retinal ischemia, may be asymptomatic
 - Abnormal DWI-MRI strongly correlated with major cause of stroke (even when neurologically asymptomatic)


Adapted from Drs. Nancy Newman and Biousse; 2015

DWI in Acute Retinal TIA/Ischemia

- DWI-MRI identifies subgroup of patients at very high risk of major stroke
- DWI-MRI needs to be performed within 24/48 hours of visual loss to allow for effective prevention of recurrent stroke

Adapted from Drs. Nancy Newman and Biousse; 2015

Tell the patient:

- "Go to the Emergency Department"
-  "Tell them you had a retinal stroke"
- Do not send these patients to their PCP, cardiologist, neurologist, neuro-ophthalmologist
- Do not try to obtain the workup yourself

Adapted from Drs. Nancy Newman and Biousse; 2015

ODE TO AN ARTERY OCCLUSION

When the vision is poor and the fundus is pale,
A branch or laminar emboli has caused the fail.
Heroic measures are rarely helpful,
And vision return is doubtful.
In an Oldie, always remember giant cell it may be.
Hurry and get an ESR and CRP.
The retina is infarcted and dead,
So neo you should not dread.
But here is where you must not choke,
Send them to the ER because they are having a stroke

Joseph Sowka, OD