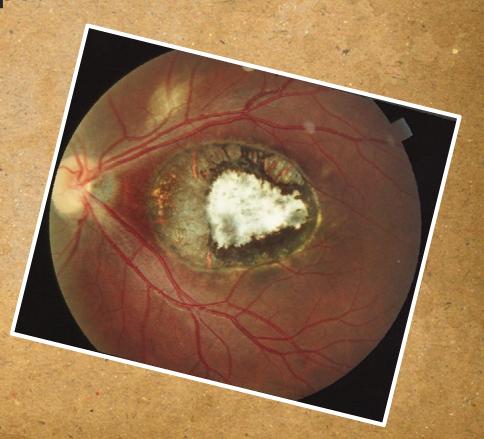
Grand Rounds

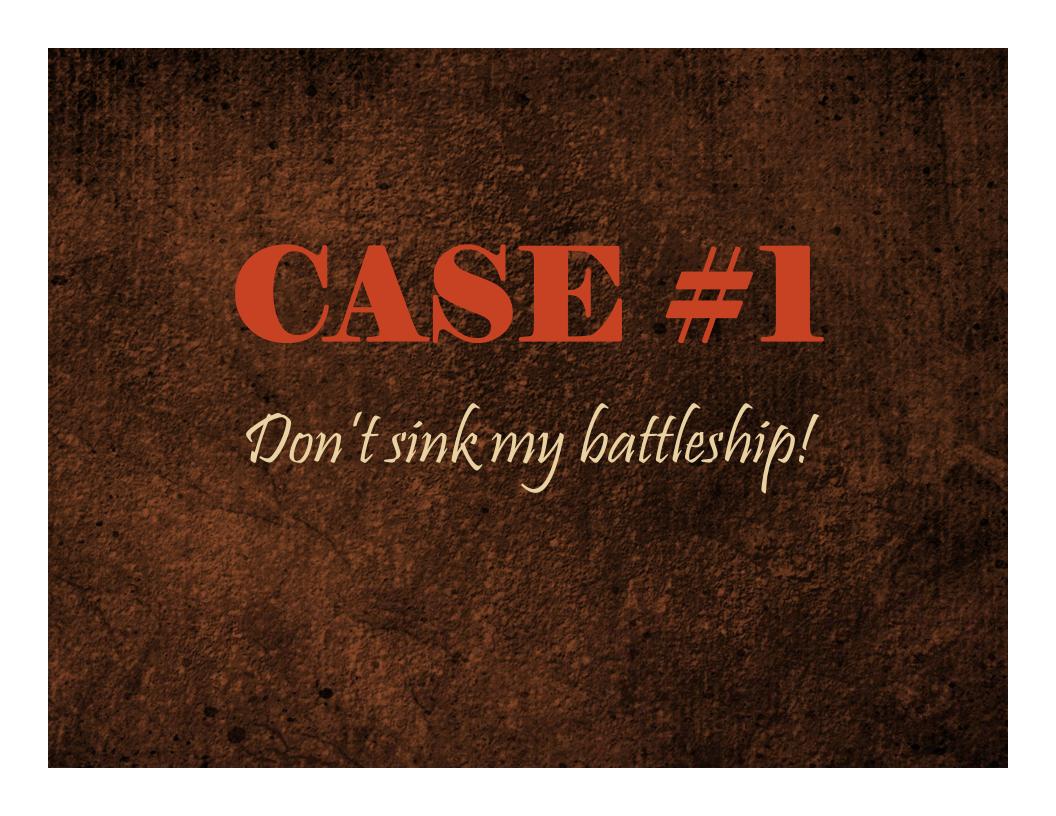
POSTERIOR SEGMENT

Richard Trevino, OD, FAAO Rosenberg School of Optometry University of the Incarnate Word

Grand Rounds: Posterior Seg

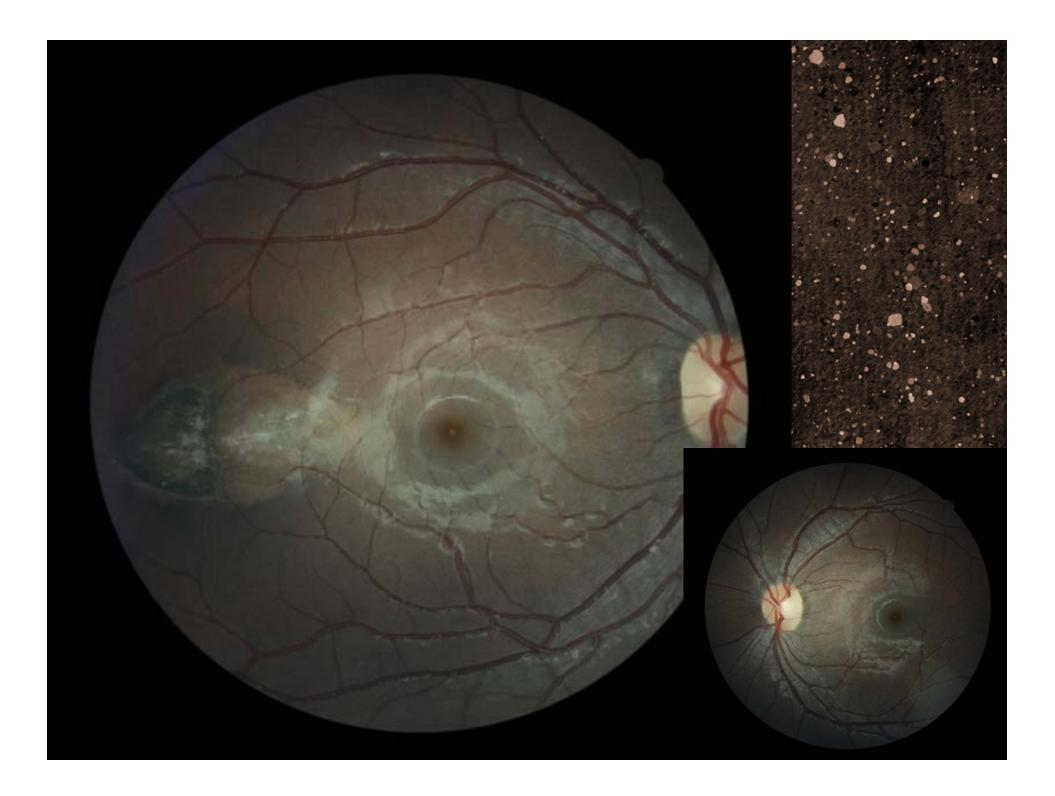
- Online slides
 - slideshare.net/rhodopsin
- Online notes
 - richardtrevino.net
- Email me
 - rctrevin@uiwtx.edu
- Disclosures
 - None

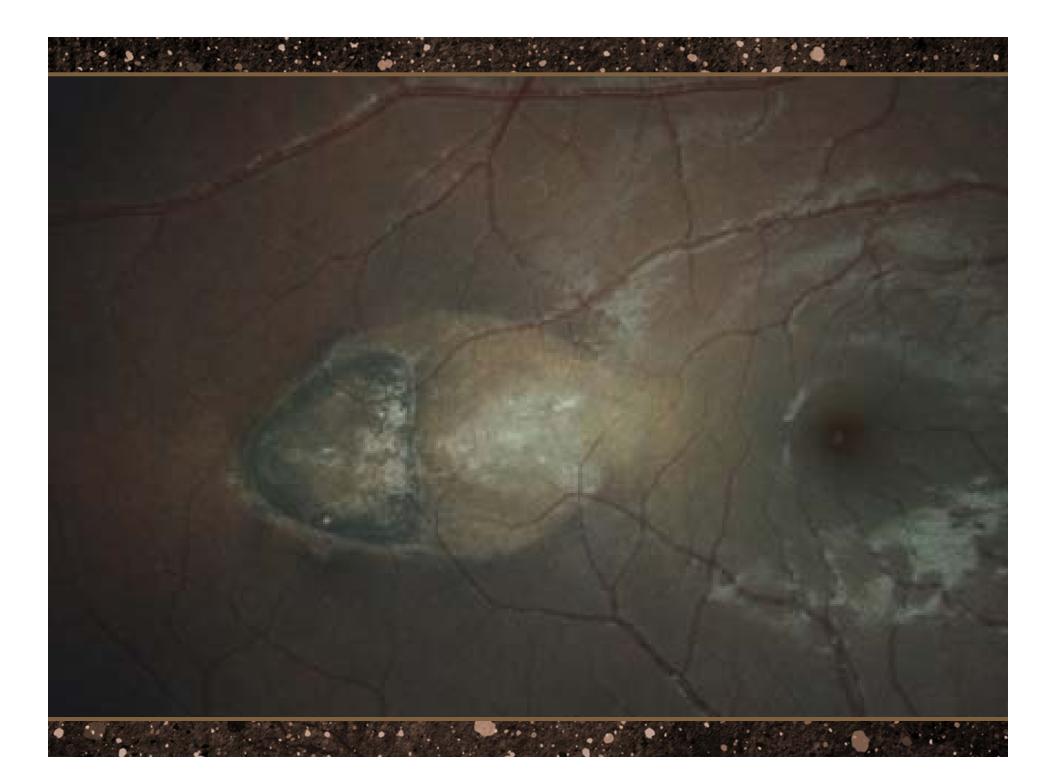


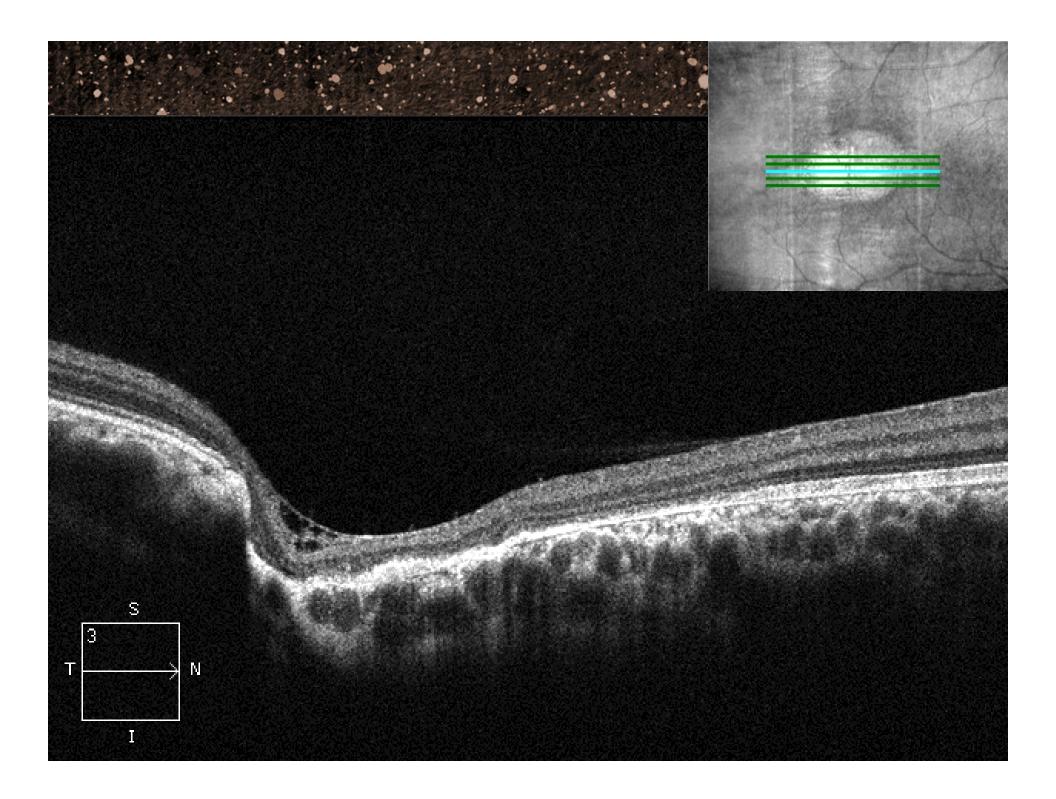


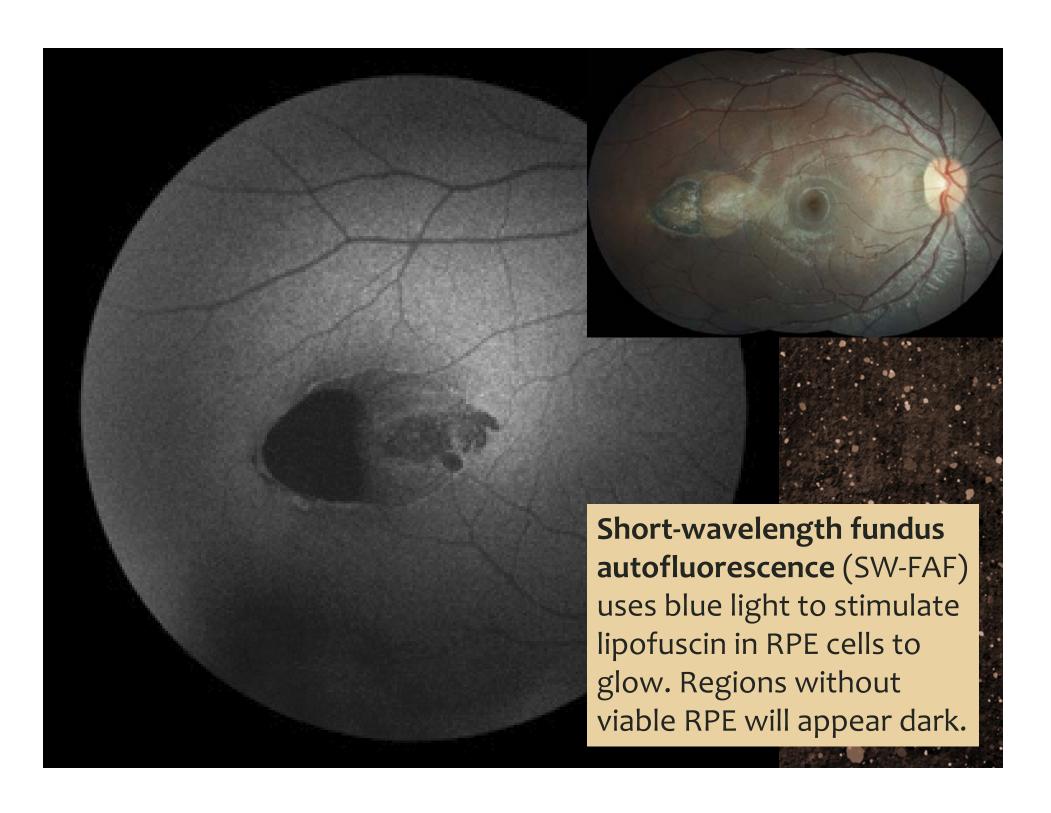
CASE #1

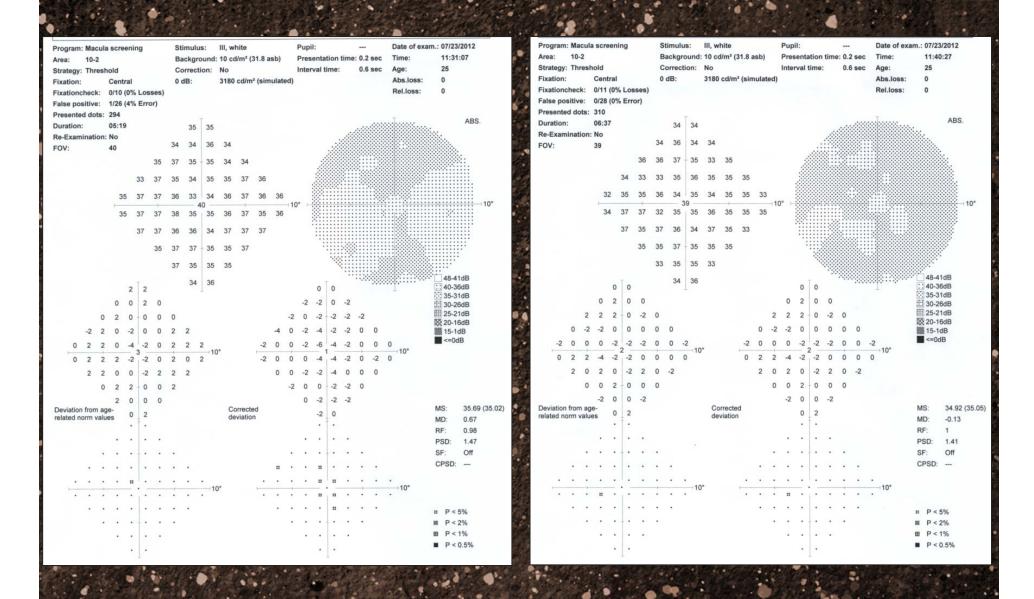
- 25yo East Indian woman presents without complaints for routine exam
- POH: Unremarkable. LEE: 1yr.
- MH: Good health. No medications
- Vision: 20/15 each eye without correction
- Entrance testing: Normal
- External exam: Normal OU
- Tonometry: 14/13 @11:00AM

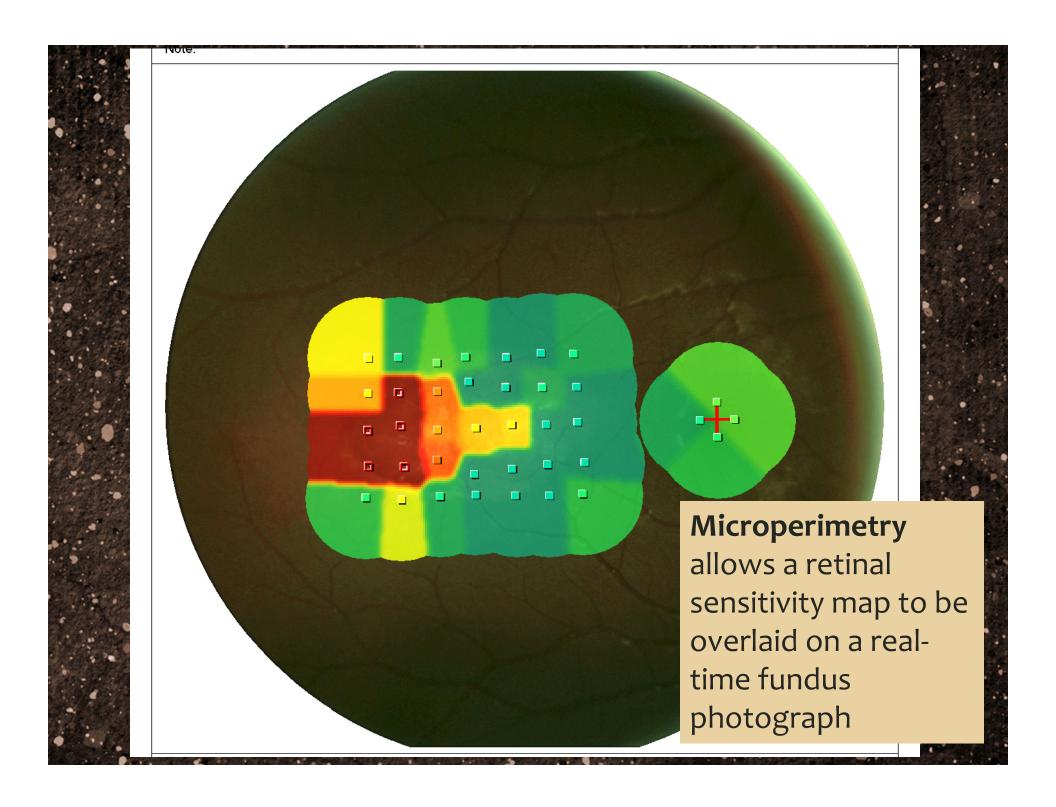










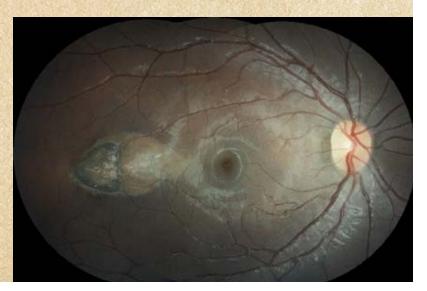


Macular coloboma (macular dysplasia)	A heterogenous group of developmental abnormalities. Often familial, frequently bilateral, systemic abnormalities not uncommon.
Macular dystrophy	Widespread retinal dysfunction, bilateral macular lesions, inheritance pattern, may be associated with features of Leber's amaurosis or RP. Electrophysiologic abnormalities
Macular scars	Intrauterine infection with Toxoplasma gondii resulting in congenital toxoplasmic chorioretinitis.
Amelanotic choroidal nevus	Shares all the features of a typical choroidal nevus minus the melanin. In general, amelanotic lesions have a better prognosis than pigmented lesions
Torpedo maculopathy	Typically unilateral congenital abnormality, characteristic "torpedo" shape, always located temporal to the fovea

...

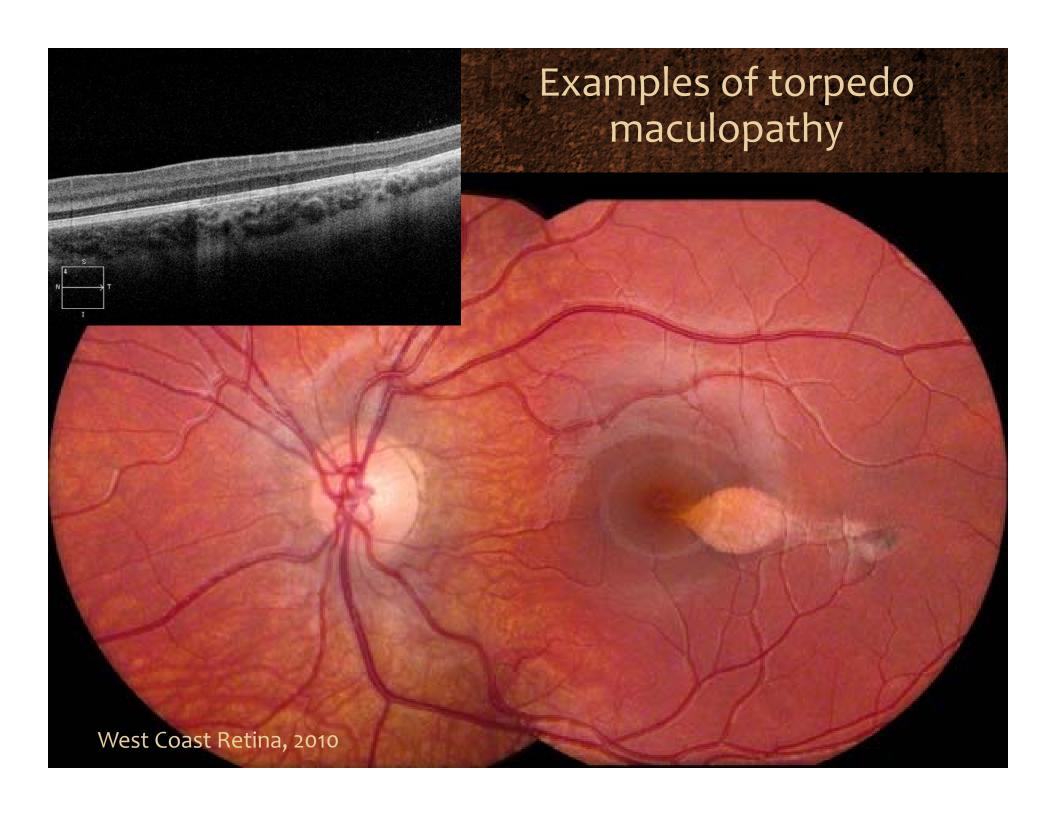
Torpedo maculopathy OD

- No specific treatment is indicated
- Patient education
- Routine eye care

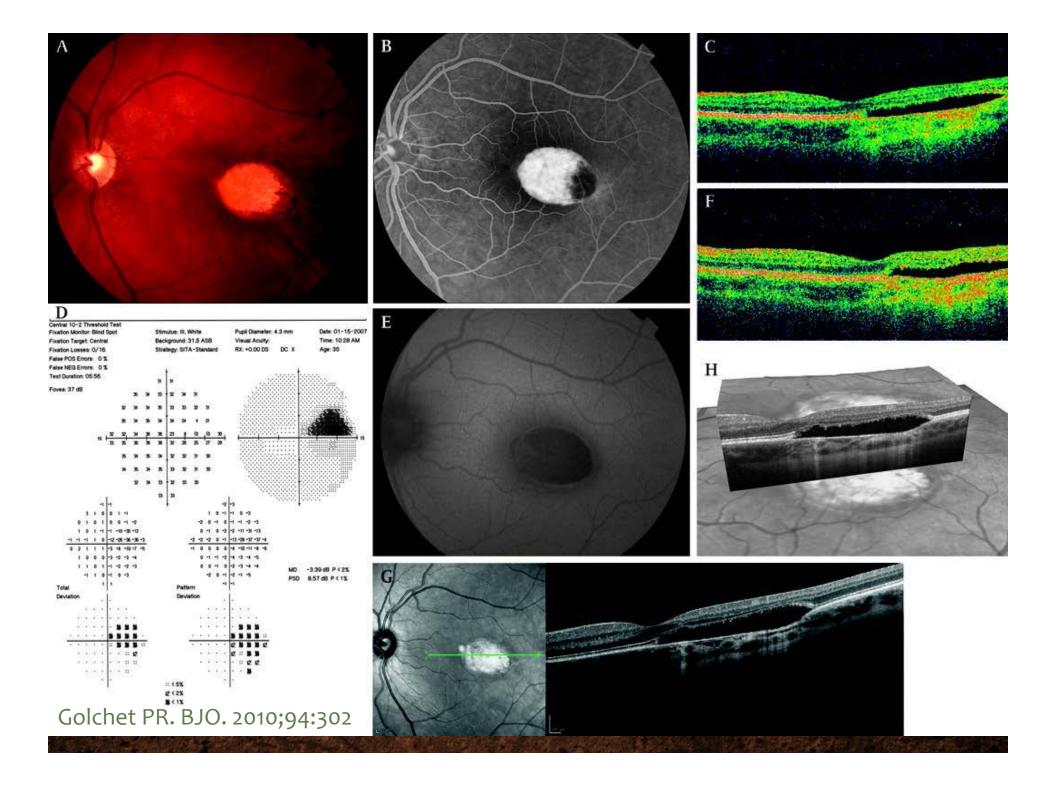


Torpedo Maculopathy

- Congenital, hypopigmented torpedo-shaped lesion in the temporal macula along the horizontal raphe
- Developmental defect of unknown etiology
- May encroach upon fovea, but rarely causes significant loss of vision
- Diagnosis based upon characteristic appearance and nonrandom location
- No treatment is required for this stable congenital lesion



Examples of torpedo maculopathy Bedar MS. Ophthalmologe, 2013;110:173



Torpedo Maculopathy

Range of clinical presentations

Constant features

- Congenital
- Location temporal to fovea
- Horizontally oval shape
- Hypopigmentation

Variable features

- Schisis cavity
- Excavation
- Degree of pigmentation
- Visual defect



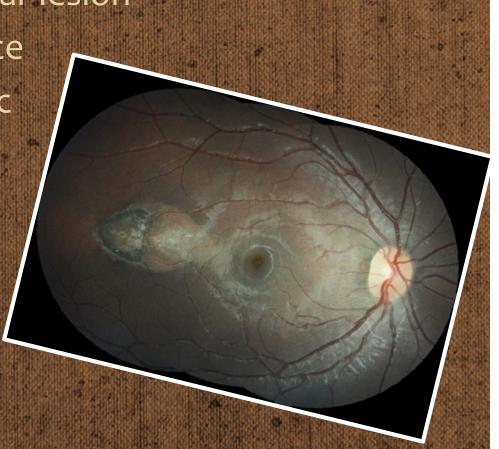


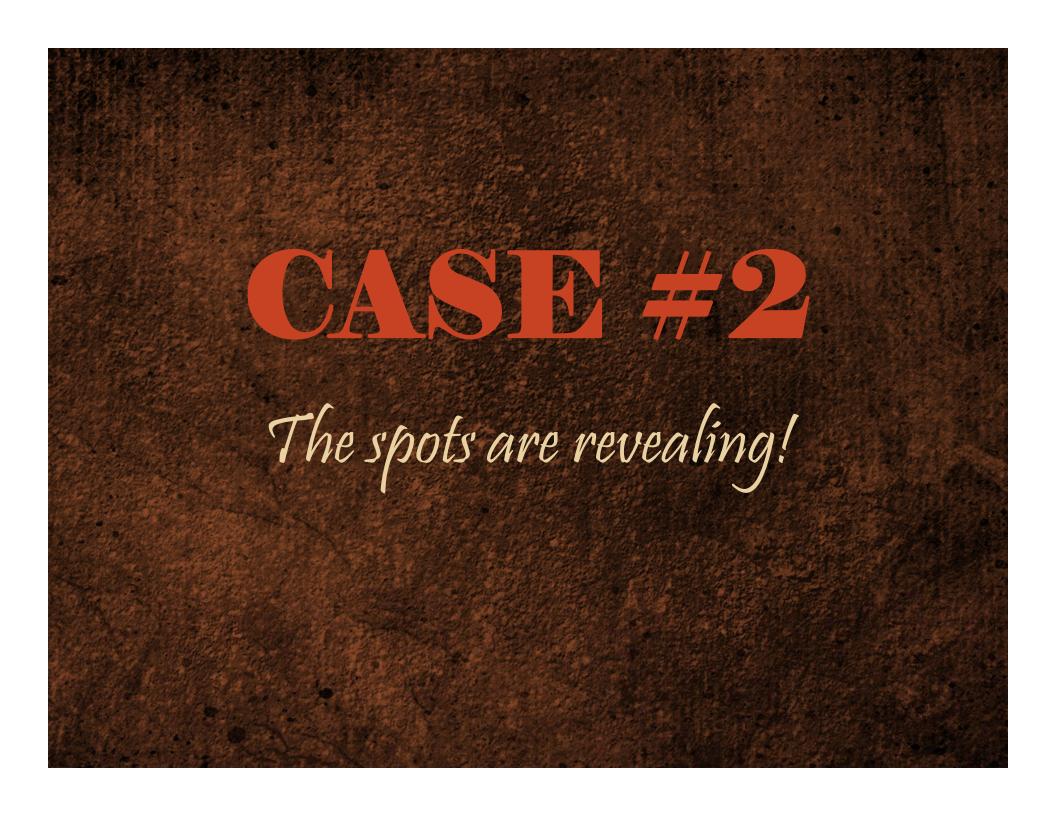
Congenital perimacular lesion

Distinctive appearance

Usually asymptomatic

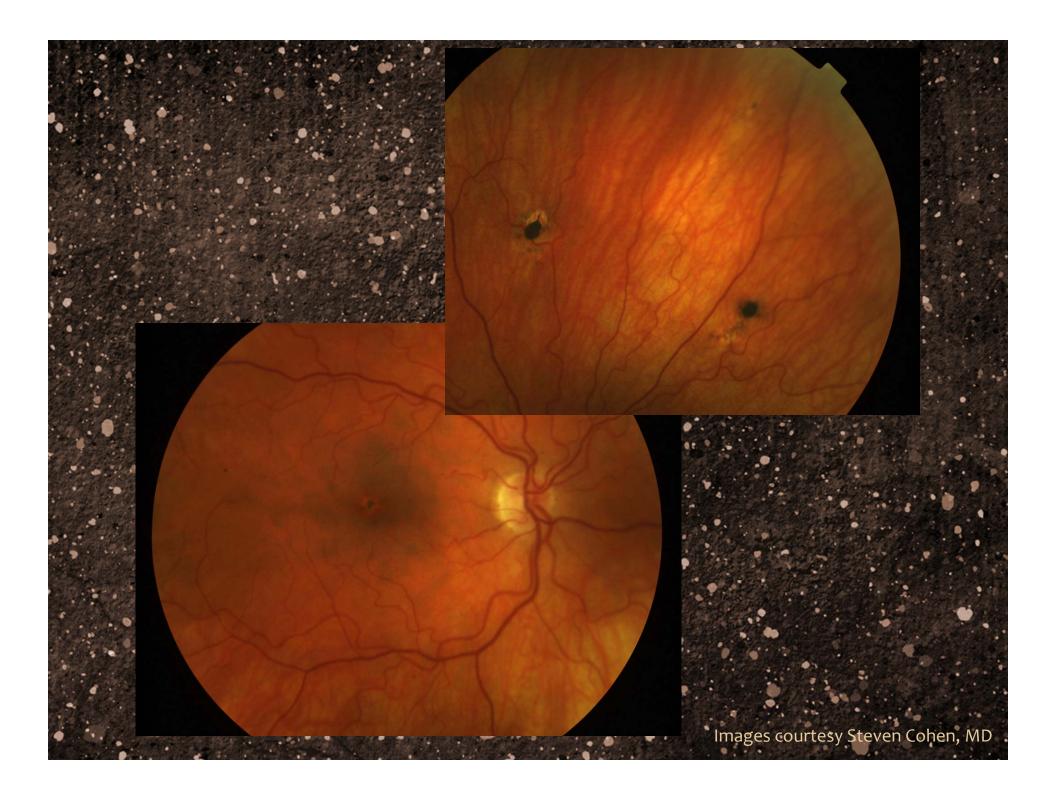
Requires no special management





Case #2

- Parents bring 7yo child in for his first eye exam. No complaints
- POH: No h/o any eye problems
- MH: Good health. No meds
- FOH: Unremarkable
- Vision: 20/20 each eye without correction
- Pupils and motility: Normal.
- IOP: 12/13 mmHg @ 9am
- External: Normal





Presumed ocular histoplasmosis	Clinical triad of multifocal chorioretinal scars, peripapillary scarring and exudative maculopathy.
Grouped pigmentation	Multifocal CHRPE lesions ("bear tracks") are typically unilateral and clustered in one quadrant of the eye.
Hamartomas of the RPE	Congenital, noncancerous tumor-like malformations of the RPE. Lesions are typically heavily pigmented, small, well-circumscribed, and may be slightly elevated
CHRPE	Congenital, flat, pigmented, well defined fundus lesion composed of intensely pigmented, hypertrophied RPE cells. Typically unilateral and solitary.

- Multiple, bilateral CHRPE-like lesions
- Suspect familial adenomatous polyposis

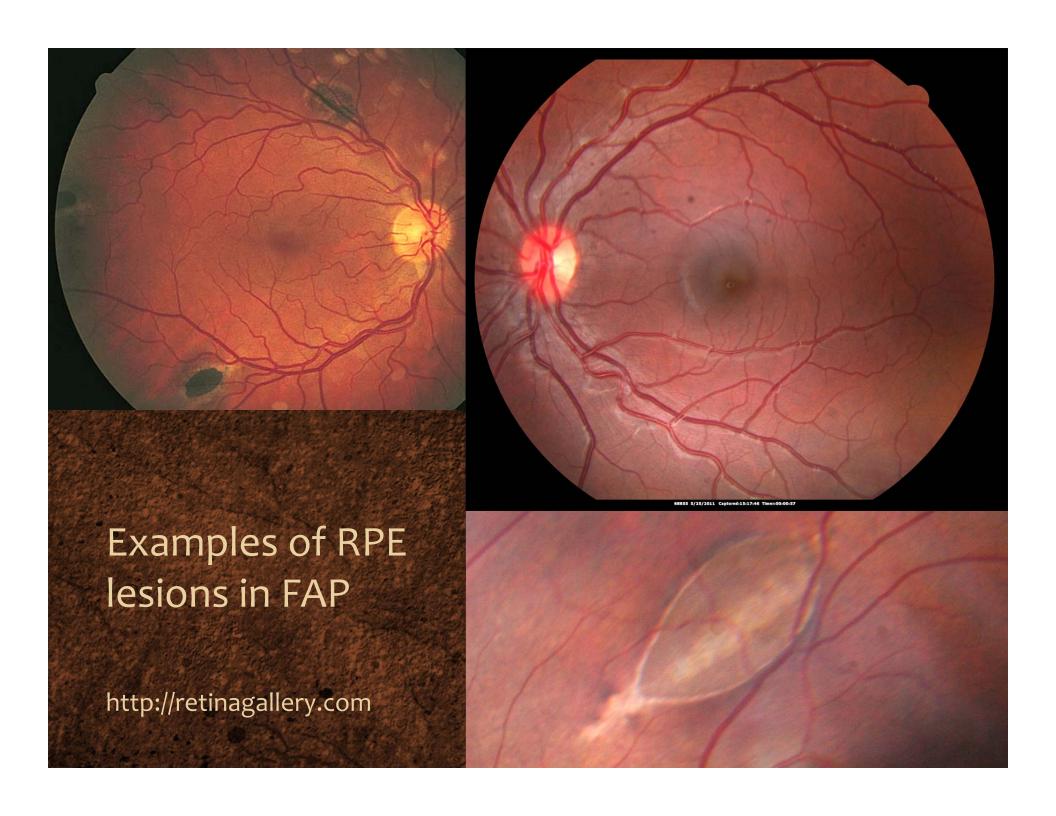
- Gastroenterology consult negative colonoscopy of child and parents
- Genetic testing offered declined by parents
- Medical surveillance for onset of polyposis

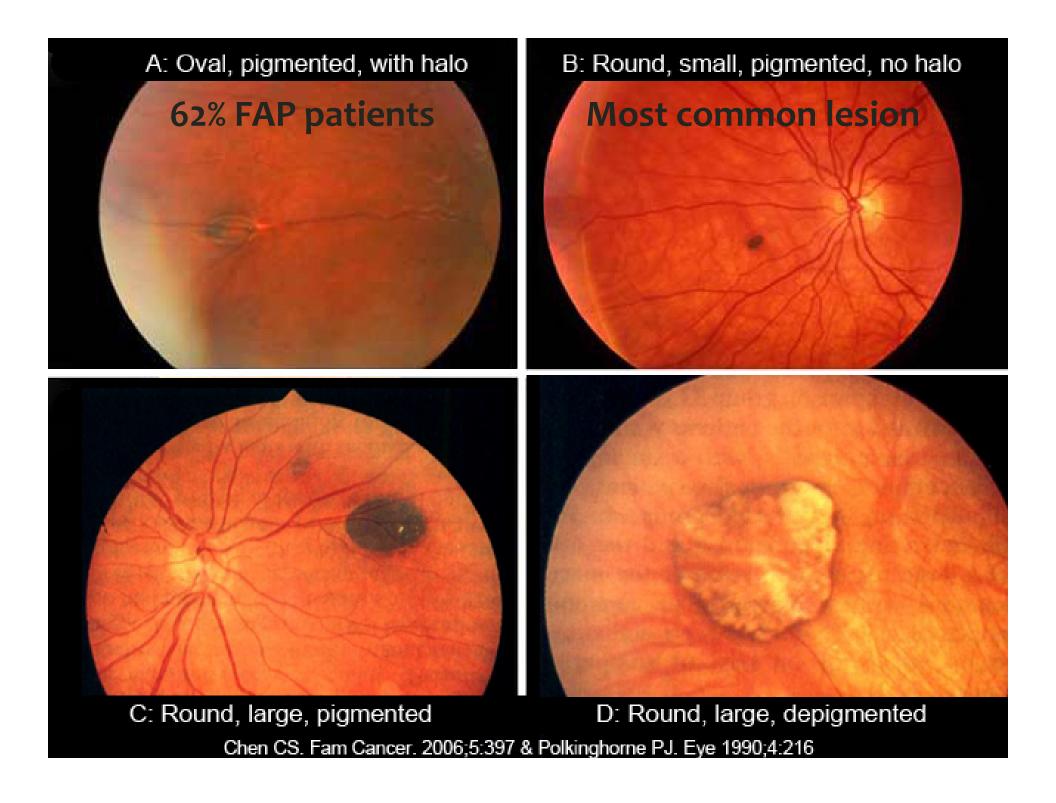
Familial Adenomatous Polyposis

- An uncommon hereditary form of colon cancer (autosomal dominant)
 - About 1% of all colon cancer in US annually
- 20% of cases have no FH of FAP, suggesting spontaneous mutation of the disease gene
- Certain FAP genotypes are associated with CHRPE-like retinal lesions (hamartomas)
 - Retinal lesions are a specific and reliable clinical marker for FAP in these patients

CHRPE-like Lesions in FAP

- Gardner syndrome: Extracolonic manifestations in patients with FAP
- Clinical appearance similar to traditional CHRPE
- Average number of lesions is 6 lesions/eye
- Bilateral in 86% of cases
- Most lesion are < 0.5 DD in size. Some patients will have numerous pinpoint lesions
- Larger lesions tend to be more oval or "tear drop" shape with a hypopigmented tail





Retinal Lesions as a Genetic Marker

- Retinal lesions are a sensitive and specific marker for FAP mutation carrier status in those families with retinal lesions (70%)
- Congenital retinal lesions may serve as an early marker for those patients destined to develop polyposis later in life
 - Onset of polyposis: age 25 yrs



CHRPE-like lesions may signal risk of colon cancer

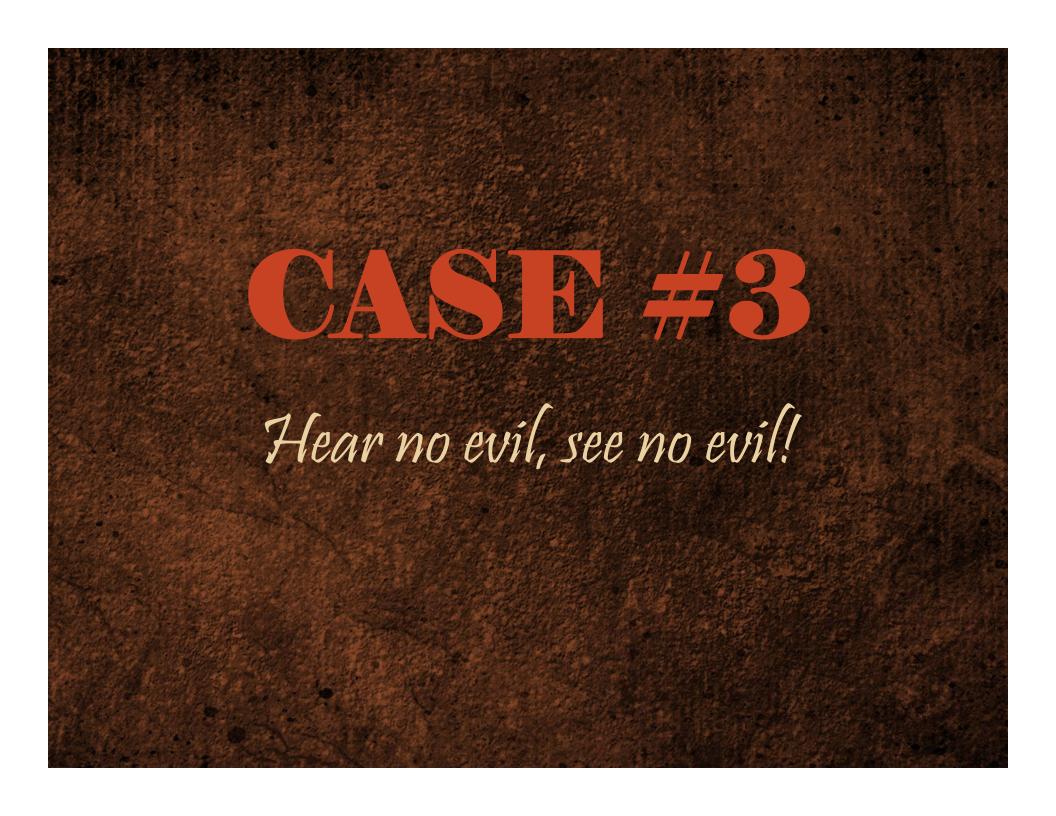
How to spot suspicious CHRPE-like lesions

1. Bilaterality

2. > 3 lesions per eye

3. Lesions associated with a depigmented streak



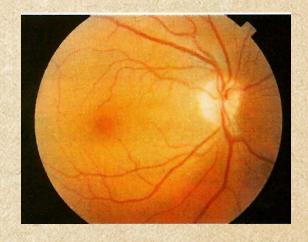


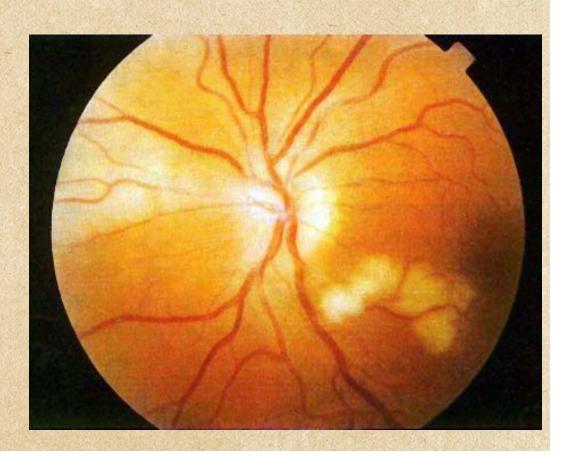
Case #3 - 1993 - Visit 1

- 36yo WF presents with c/o "gray haze" OS x
 2 weeks
 - 1 week prior had suffered left-sided HA and photopsias OS that sent her to the ER. Exam was normal and Fiorinal with codeine was prescribed
- POH: Normal.
- MH: Head and neck aches and parestheias in her arms following MVA x 2yrs
- FOH: Father with glaucoma

Case #3 - 1993 - Visit 1

- Vision 20/20 in each eye
- Pupils: Normal
- Motility: Normal
- IOP: 13 mmHg OU
- External: Normal





- Acute BRAO OS
- Evidence of older resolving BRAO OS

- Carotid duplex scan Normal
- Echocardiography Normal
- Labs: CBC, rheumatoid factor, RPR, fasting glucose, ANA – all normal
- Start ASA
- Retinal lesions resolved @ 1 month F/U visit

Case #3 - 1996 - Visit 2

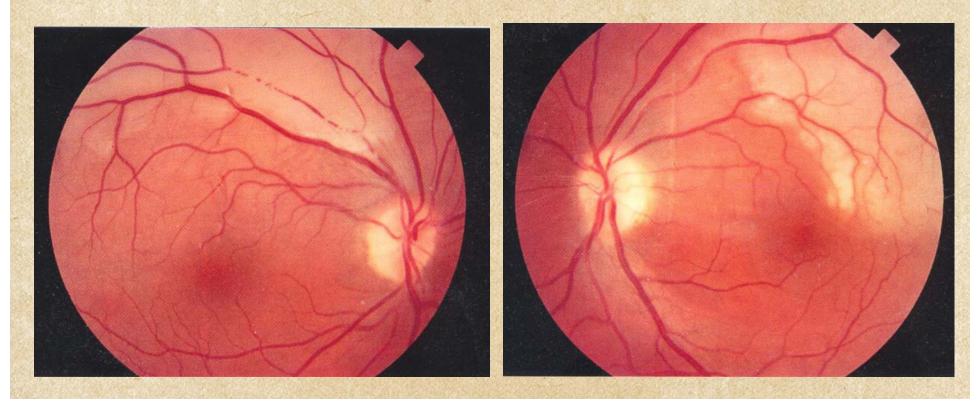
- C/O constant "flash bulb glare" OS x 1 day
- Vision: 20/20 OD, 20/25 OS
- Pupils normal, Motility normal, IOP 14/12 mmHg
- External: Normal OU
- DFE: Normal OD, BRAO of superior-temporal artery OS
 - No visible embolus
 - Occlusion does not appear to occur at bifurcation

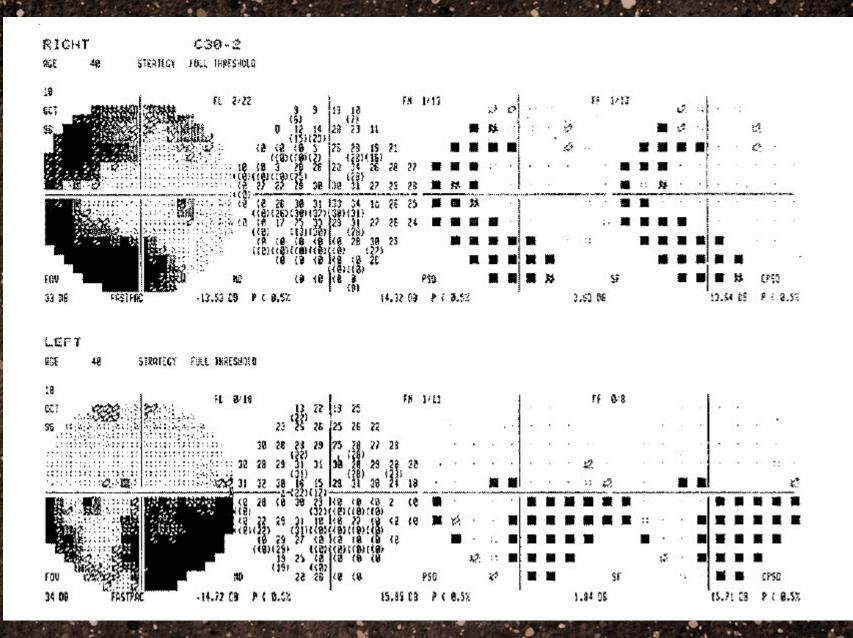
BRAO OS

- Retinal consult with FA –diagnosis of idiopathic recurrent BRAO. No known cause or tx for condition
- IM consult: Obese (238lbs) but otherwise in good health. No BCPs, no vasculitis, synovitis, diabetes, or HTN
- Continue ASA, start low fat diet

Case #3 - 1996 - Visit 3

- Presented 2 weeks later with photopsia OD
- Examination was remarkable for the presence of new BRAO OD





Assessment

Idiopathic recurrent BRAO, OU

Management

- IM consult: Negative evaluation except mildly elevated ESR (27 mm/h, normal: 0-20)
- Rheumatology consult: Negative evaluation.
 Normal temporal artery biopsy
- Audiometry and otolaryngology consult: moderately severe sensorineural hearing loss. Referred for hearing aid fitting

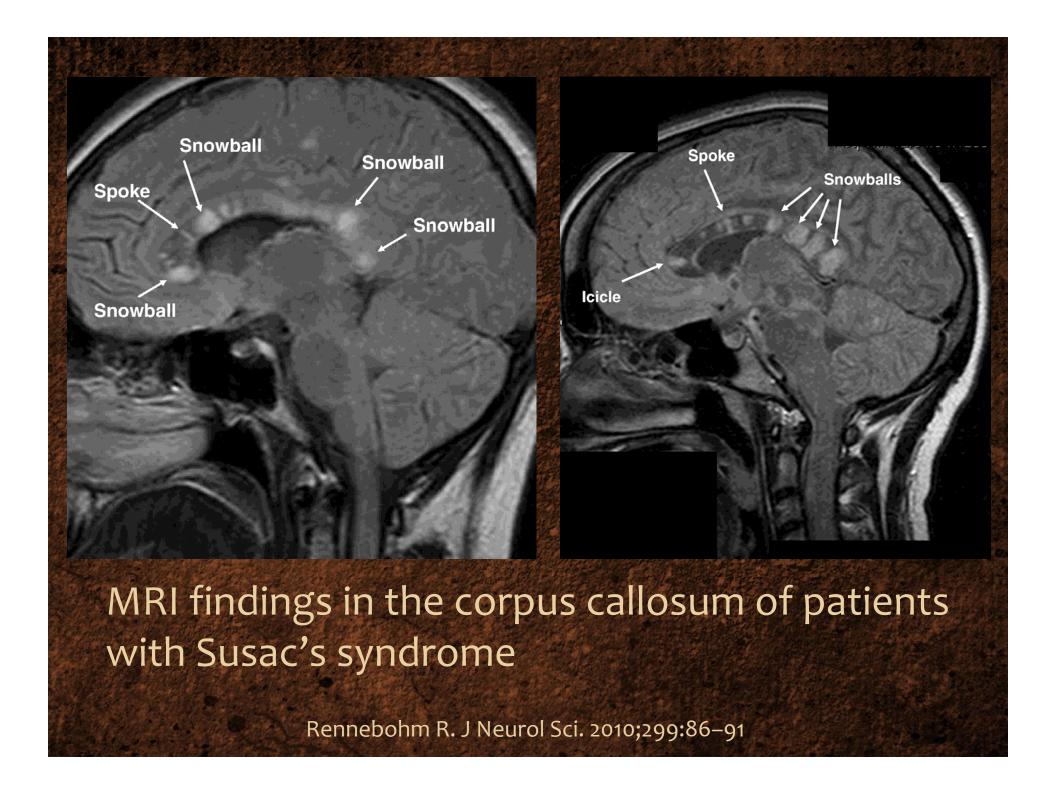
Multiple sclerosis	Uveitis and retinal phlebitis are among the posterior segment manifestations of MS. BRAO is not associated with MS	
Systemic lupus erythematosis	Retinopathy is frequently associated with SLE may include CWS, BRAO, neovascularization and VH. Diagnosis made on clinical and lab grounds.	
Sarcoid	Antiphospholipid antibody syndrome may be associated with sarcoidosis and can lead to retinal artery occlusion	
Susac's syndrome	Clinical triad of encephalopathy, BRAO and hearing loss	
Lyme disease	BRAO has been reported as an uncommon complication of ocular Lyme borreliosis	

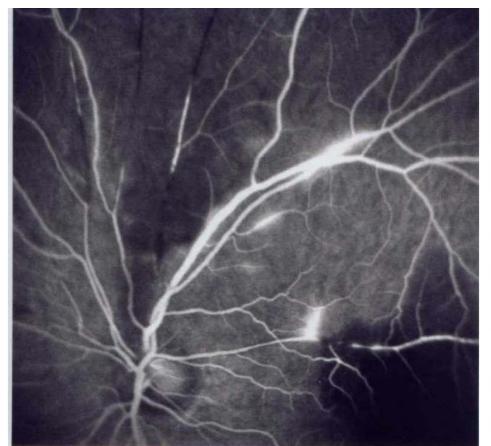
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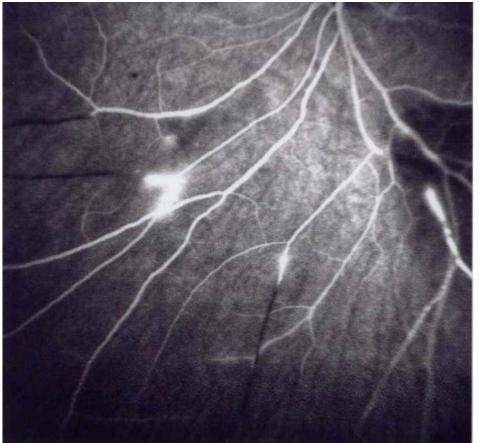
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Susac's Syndrome

- Clinical triad of multifocal encepholopathy, BRAO, and hearing loss that typically occurs in young adult women
- First described by Susac in 1994
- Etiology believed to be immune mediated endotheliopathy affecting blood vessels in the retina, chochlea and brain
- Diagnosis aided by MRI findings of "snowball" lesions in the corpus callosum
- Treatment with steroids and immune suppressants can slow progression of the disease









Distinctive findings include arterial wall hyperfluorescence on FA and so-called "Gass plaques" – refractile yellow deposits believed to represent focal damage to the arterial wall

Egan RA. J Neurol Sci. 2010;299:97–100

BRAO in the Young

- Associated with predisposing conditions that are different from those found in older patients with BRAO
- Women affected twice as frequently as men when trauma-associated occlusions are excluded
 - Hormonal influences, pregnancy, oral contraceptives

BRAO in the Young

Emboli	Cardiac disease, IV drug abuse
Thrombosis	Pregnancy, BCP use, Coagulopathy
Arteritis	Lupus, Lyme
Arterial spasm	Migraine, Drug abuse (cocaine, meth)
Vascular compromise	Orbital, optic nerve, retinal disease; Trauma

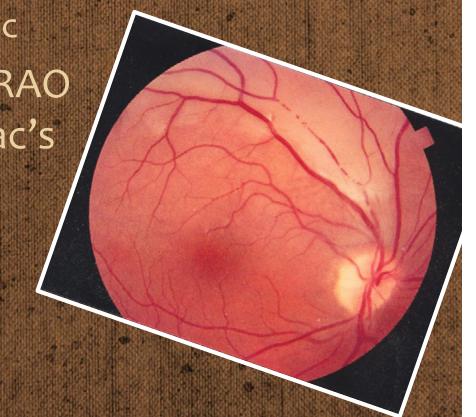
Take Home Message

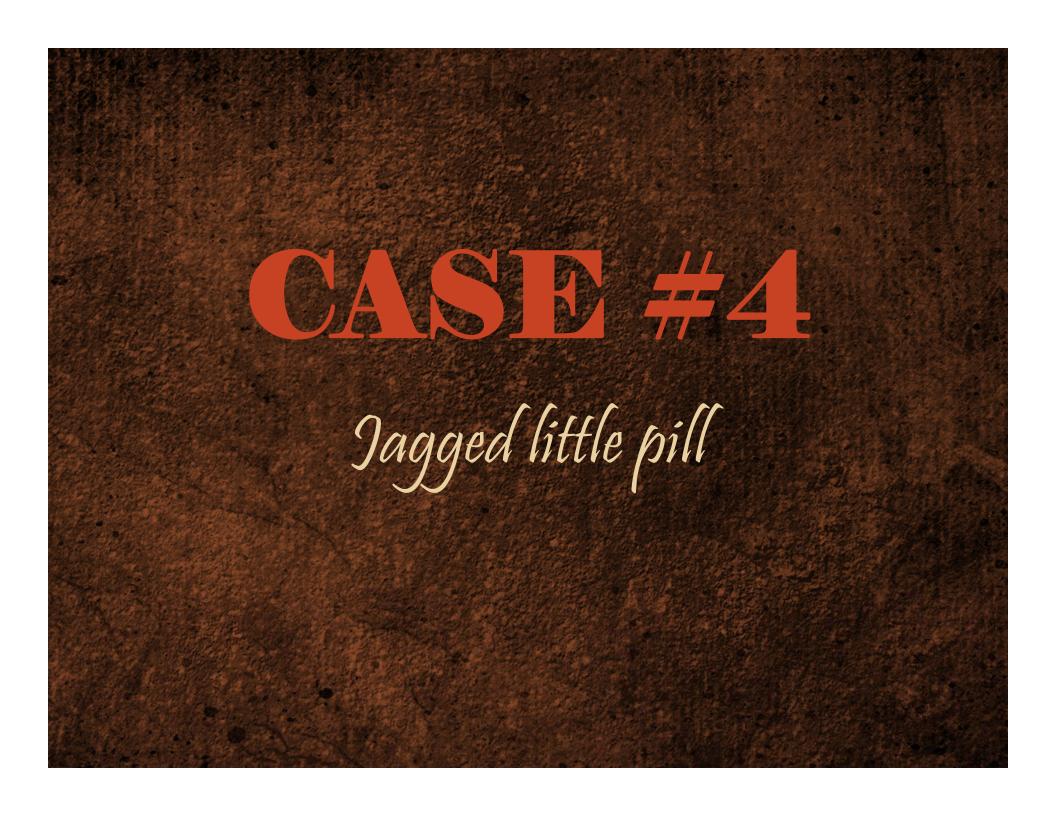
 The causes of BRAO in young people are different than in older people

Less likely to be embolic

 Idiopathic recurrent BRAO may be caused by Susac's syndrome

Check for hearing loss and MRI lesions



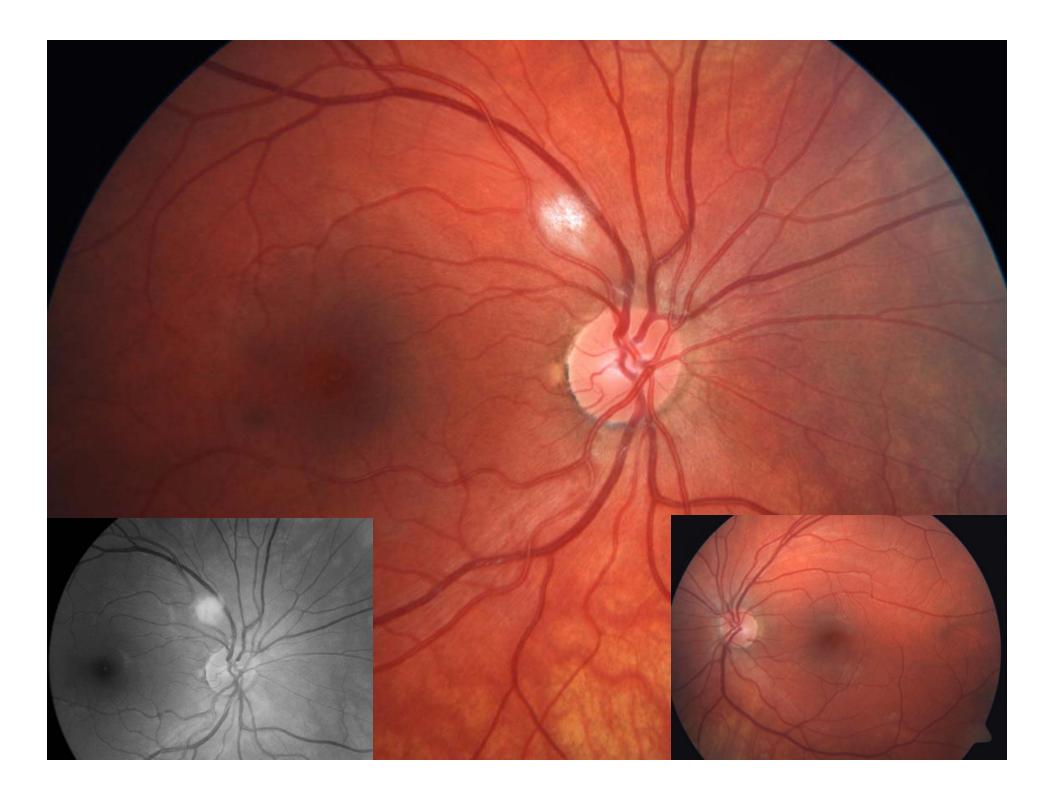


Case #4

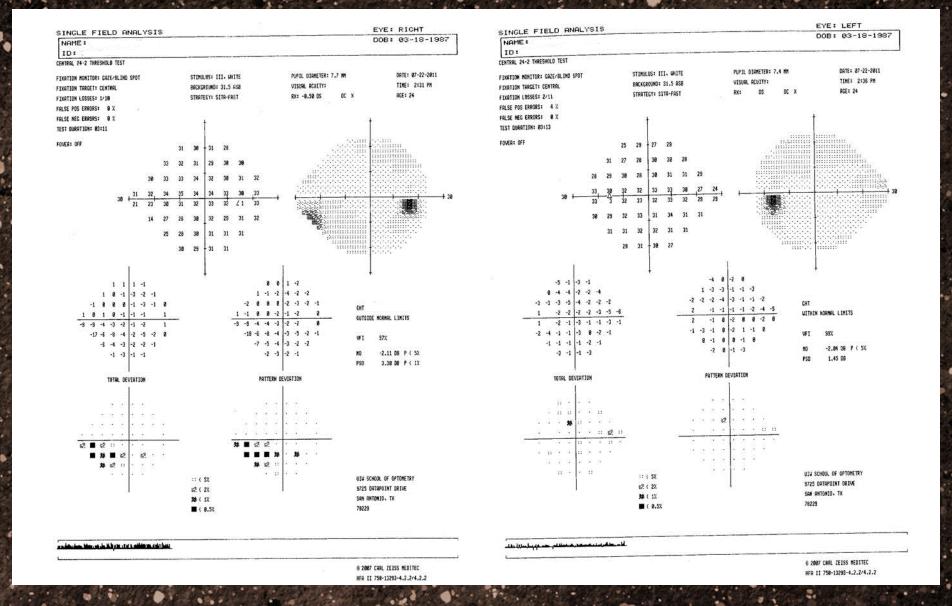
- 24yo WF presents with c/o "blurry light spot" in the *inferior-nasal* paracentral vision of the right eye x 1 week. Symptoms are made worse while exercising.
- POH: LEE 2yrs. Negative for any prior eye dx
- MH: Good health. Nonsmoker. Meds: BCP
- FH: MGM with diabetes

Case #4

- BVA: 20/20 in each eye
- Pupils and motility: Normal
- BP: 113/71 RAS, Pulse: 81 bpm
- IOP: 13/12 mmHg @ 4pm
- Amsler: Blurred region <u>inferior-temporal</u> to fixation OD, Normal OS
- Color: Normal OU (HRR)
- External: Normal OU







Assessment

 Isolated cotton wool spot in an apparently healthy young woman

Management

- Medical evaluation to identify cause for CWS
- Recommend D/C BCP and start ASA
- Follow-up in 2 weeks

Idiopathic CWS	It has been reported that an underlying disorder can be found in 95% of isolated CWS	
Diabetes	Undiagnosed diabetes is the most common cause of isolated CWS in an apparently normal patient	
HIV	CWS are a prominent feature of HIV noninfectious retinopathy, the prevalence of which is inversely related to the patient's CD4+ count	
Lyme disease	Cotton wool spots have been reported as a sign of Lyme retinitis	
BRAO	CWS are a universal feature of BRAO	

Medical Evaluation

- Physical exam by PCP was normal
- Normal laboratory testing: Fasting glucose, CBC, ANA, Rheumatoid factor, C-reactive protein, HIV screen
- Normal carotid Doppler and echocardiogram
- FTA-ABS was minimally reactive
 - Serologic ELISA testing for Lyme disease recommended
- Follow-up: Photopsia persisted x 4-6 wks before abating. VF defect remained unchanged

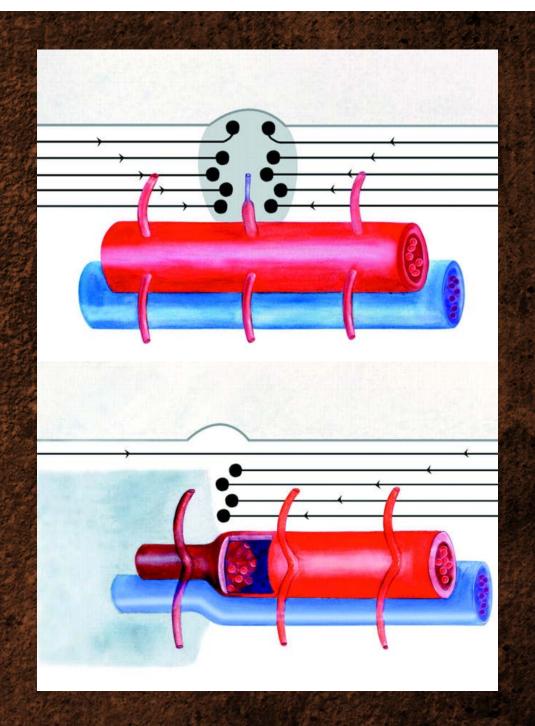
What is a CWS?

- CWS is a localized accumulations of axoplasmic material within adjacent bundles of ganglion cell axons.
- Two clinical presentations:
 - Focal ischemia from terminal arteriolar occlusion.
 - 2. Appearance at the boundary of an ischemic region of the retina

Focal ischemia:
Occlusion of a terminal branch of a retinal arteriole (red) results in a small area of infarction (grey) in the RNFL wherein both orthograde and retrograde axoplasmic transport is obstructed.

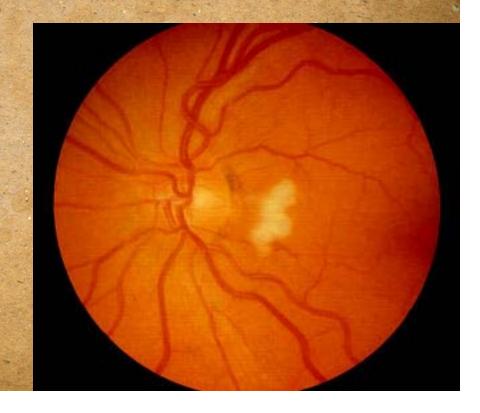
Sentinal lesion:
Occlusion of a retinal arteriole results in retinal infarction

McLeod D. BJO. 2005;89:229



Isolated CWS

- Retinal whitening associated with arteriole occlusion will resolve in about 1-2 weeks
- CWS take 3-6 weeks to resolve
- CWS will appear to be "isolated" for most of the time before they resolve



Vision Loss Associated with CWS

- CWS are almost always asymptomatic
- May be associated with localized or arcuate scotomas

OCT studies reveal permanent damage to the

inner retinal layers and RNFL at the site of a resolved CWS lesion

 Some visual recovery may occur following resolution of CWS lesions



Evaluation of Idiopathic CWS

- Directed toward those conditions that predispose the patient toward embolism and thrombosis
- Common: diabetes, hypertension, and collagen vascular disease
- Less common: HIV and other infections, hematologic disease and coagulopathies, pancreatitis, embolic disease, trauma, pregnancy, and idiopathic conditions

OCP and Thromboembolism

 Use of OCP increases the risk of venous thromboembolism (VTE) by about 5x

Risk is further increased in women who smoke,
 are obese, have HTN, coagulopathies, and a FH

of thromboembolic disease

Third generation OCPs

 Lower risk of MI, CVA, and other side effects (weight gain, acne, headaches and unwanted hair growth)

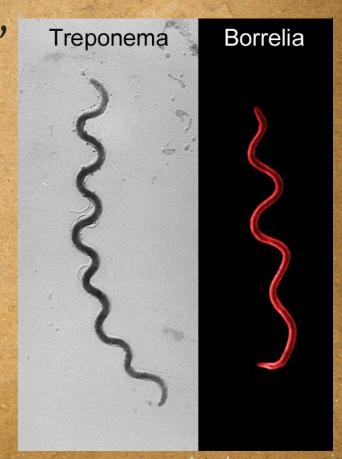
Higher risk of VTE

False Positive FTA-ABS in Lyme

 Lyme disease is caused by Borrelia burgdorferi, a spirochete that is biologically similar to the

spirochete that causes syphilis, Treponema pallidum

- This similarity is responsible for cross-reactivity of the FTA-ABS serologic test for syphillis and Lyme disease
- Some patients with Lyme disease will test positive for syphilis on the FTA-ABS test



CWS and Lyme Disease

- Many ocular manifestations of Lyme disease have been reported
- The most common appear to be conjunctivitis, uveitis (especially pars planitis), a retinopathy consisting of macular edema and vasculitis, and cranial nerve palsys
- Cotton wool spots have been reported as a sign of Lyme retinitis

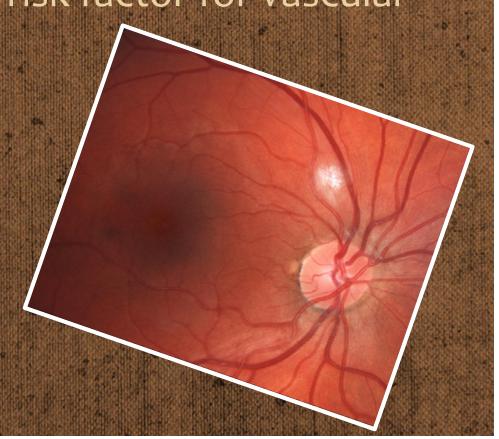
Take Home Message

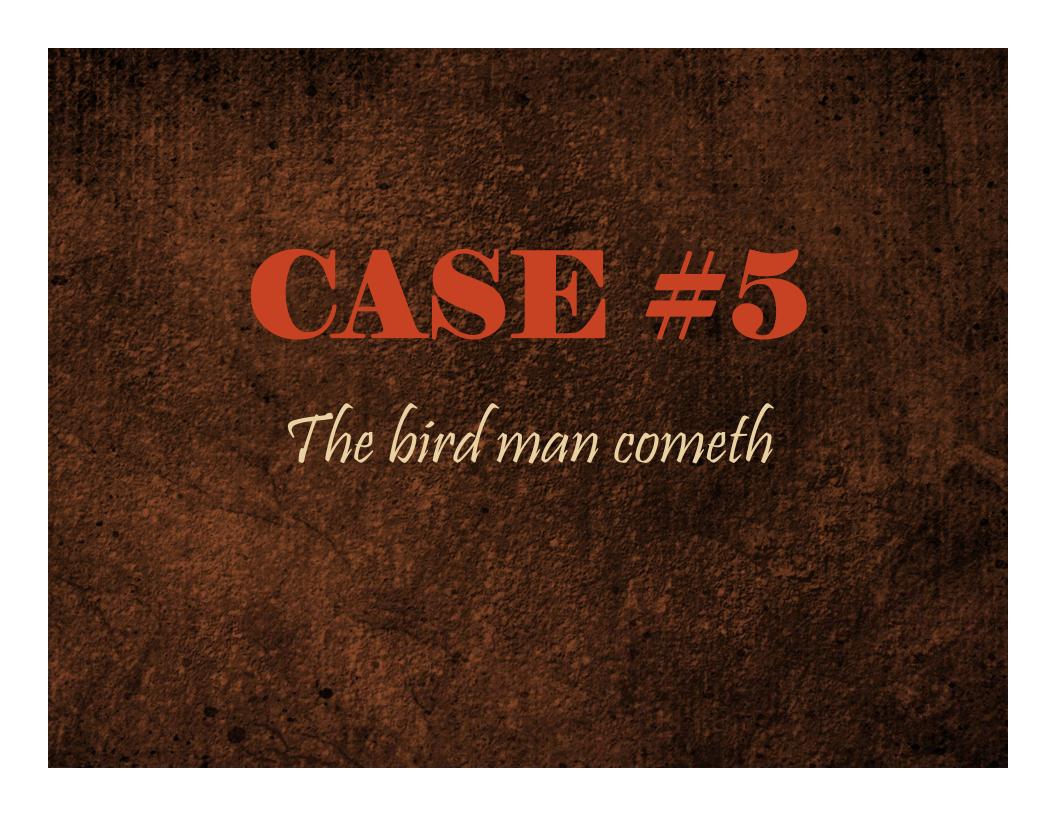
Isolated CWS are not isolated

OCP are a significant risk factor for vascular

occlusions in young healthy women

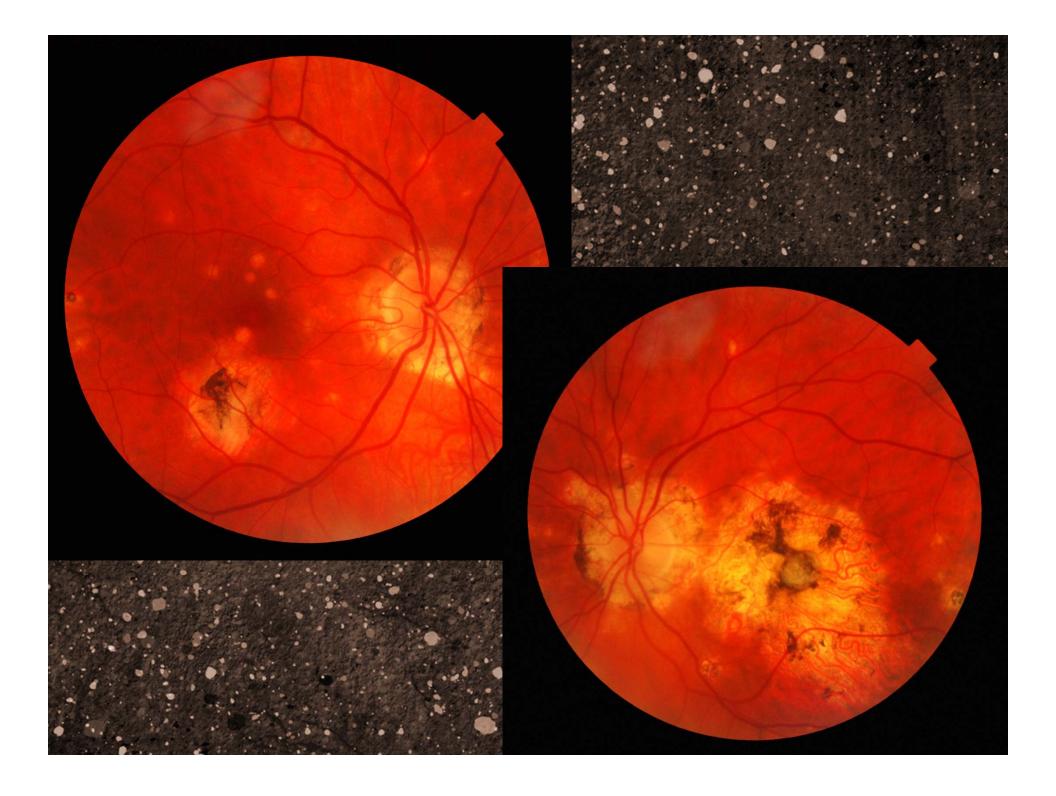
 Lyme disease may cause CWS and produce a falsepositive FTA-ABS





Case #5

- 56yo WM presents for routine eye exam
- Occupation: Maintenance man and farmer
- POH: H/O vision loss OS due to "bleeding" 5 years ago. Treated with laser
- MH: NIDDM x 4yrs (HbA1c: 6.5)
- BVA: 20/20 OD, FC @ 10ft OS
- Pupils and motility: Normal
- IOP: 14/16 mmHg @ 2pm
- External: Normal OU



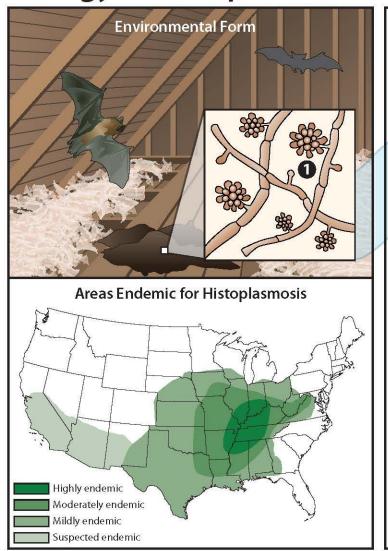
Assessment

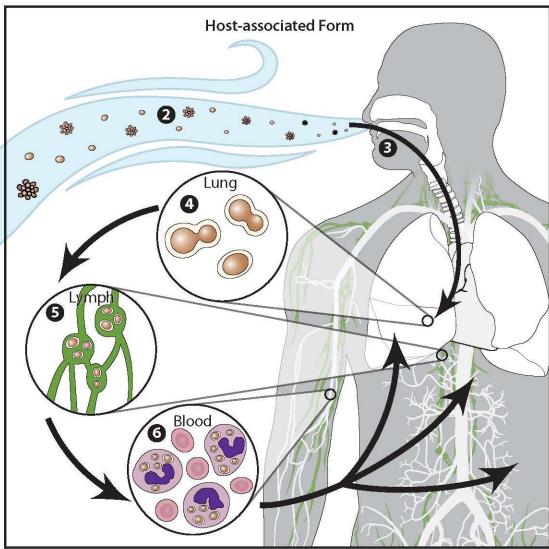
- POHS OU with maculopathy OS
- No diabetic eye disease

Management

- Recommend use of protective devices because is frequently exposed to soil and bird droppings as a farmer and maintenance man.
- Daily Amsler grid and environmental Amsler
- Safety issues for monocular patients
- Patient education diabetic eye disease.
- RTC 6 months, sooner PRN

Biology of Histoplasmosis





In the environment, *Histoplasm capsulatum* exists as a mold (1) with aerial hyphae. The hyphae produce macroconidia and microconidia (2) spores that are aerosolized and dispersed. Microconidia are inhaled into the lungs by a susceptible host (3). The warmer temperature inside the host signals a transformation to an oval, budding yeast (4). The yeast are phagocytized by immune cells and transported to regional lymph nodes (5). From there they travel in the blood to other parts of the body (6).



POHS and Vision Loss

- Vision loss occurs secondary to exudative maculopathy caused by CNV
- Reactivation of histo spots in the macular region is believed to play a role in triggering CNV
- Patients with histo spots in the macular region, especially near the fovea, should be considered at risk for vision loss

Preventing Reactivation of POHS

- Re-exposure to histoplasmosis may play a role in reactivating retinal lesions and promoting development of maculopathy
- Avoid high risk areas where histoplasmosis levels tend to be highest
 - Caves, chicken coops, old buildings
- Protect yourself or avoid high risk activities
 - Construction and demolition, working with poultry, HVAC installation or service, farming, gardening

Preventing Reactivation of POHS

- Personal protective equipment
 - Masks and respirators
- Dust control
 - High efficiency air filters
 - Vacuum cleaning
 - Wetting contaminated soil
- Endogenous factors
 - Chronic fungal infections
 - LASIK



Take Home Message

- Patients with histo spots near the central macula are at risk for vision loss due to maculopathy
- Take steps to decrease the risk of histo spot reactivation in patients at risk for vision loss



