Lumps and Bumps and Dots – Oh My!

A Posterior Segment Review

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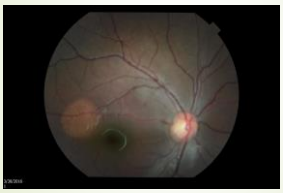
Patient here for "routine exam." We see… this!

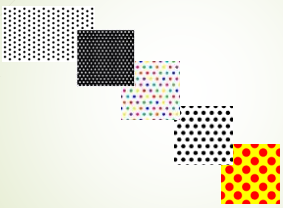
And then this…
So, let's start with: "Oh my..."



Challenges

- Chronic (Ex. Glaucoma)
  - You have time.
  - You have time to second guess yourself.
  - You have time to second guess yourself and come up with a worse decision.
  - Also action taken will naturally depend on the doctor's comfort level and the case at hand.
- Acute (Ex. Sudden onset anything!)
  - Often, little time to ponder possibilities, but…
  - Many cases will require secondary/tertiary care
  - Second guessing occurs later.
  - Patient education will take on immediacy, just like all the other aspects of care.

And On to Dots…



APMPPE

Acute Posterior Multifocal Placoid Pigment Epitheliopathy

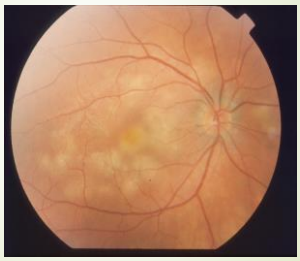
HISTORY

- 18 year-old woman
- "Black spot blocking my vision RE and several in LE."
- Neighbor's house has many bird nests; rashes on legs after an "outing."
- Recent bronchitis and UTI
- PMHx: Asthma
- Meds: Prednisone for asthma from Health Center (25 mg. tapering dose)

OCULAR EXAMINATION

VA:	OD CF at 5'	OS 20/40
Pupils:	PERRLA; no APD	
EOM:	Full	
VF defect:	OD central defect	OS superior defect
IOP:	OD 18	OS 18
SLEx:	OD NL state	OS NL state

18 year old woman; OD



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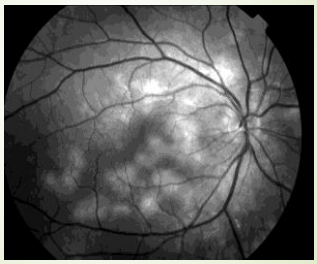

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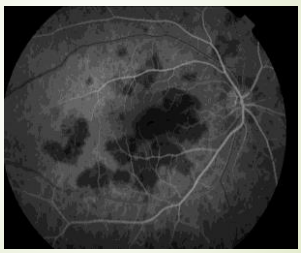

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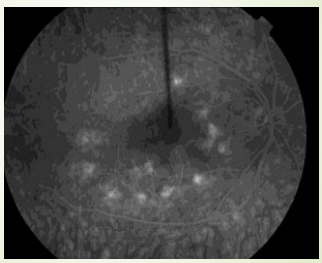

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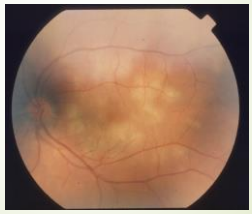

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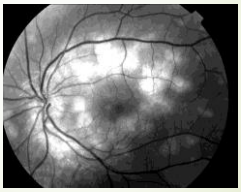

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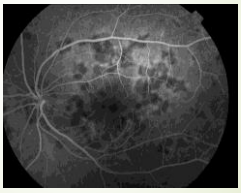

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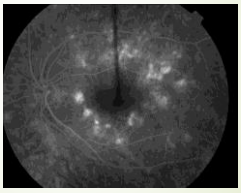
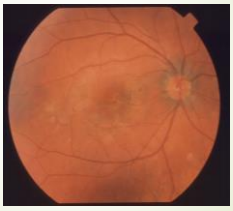


TWO WEEKS LATER

- Subjectively, "No significant change."
- VA: OD CF 5' OS 20/20
- Pupils: PERRLA
- IOP: OD 14 OS 14
- SLEx: OD NL OS NL

F/U 2 weeks; OD



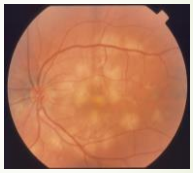
FOUR WEEK FOLLOW-UP

- New visual loss OS "for 5 days."
- "Left eye is now worse."
  - Note: OD improved in the third week; statement reflects any change after the initial improvement
- Meds: Prednisone (increased initially to 60 mg, tapering dose)

Here we go again

- VA: OD 20/25 OS 20/100
- Pupils: PERRL
- IOP: OD 15 OS 14
- SLEx: NL State OU

OS



Finally… seven weeks from initial presentation

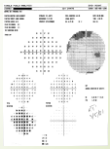
VAs 20/25 OD, OS

39-Year-Old Man

- "I've lost vision in my left eye. Comes and goes."
- Long-standing Hx of ophthalmic migraine; has never had the headache until recently
- Borderline high BP
- No medications

Physical Exam

- VA: OD 20/15 OS 20/50
- All externals: NL state OU
- VF 30-2
  - Nothing wrong!
  - Patient's imagination



Internal

- Case profile atypical.
  - Moderate vitritis noted OS
  - IOPs slightly elevated OS
  - OD NL state
  - OS Nerve-fiber layer swelling
- Patient referred to retinologist for evaluation and co-management.

One week later

- "Vision much worse."
- VA: OD 20/15 OS 20/100
- Pupils: Subtle APD OS
- Fundus evaluation:

March

March

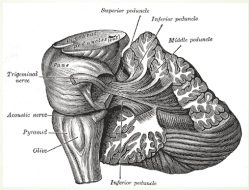
May

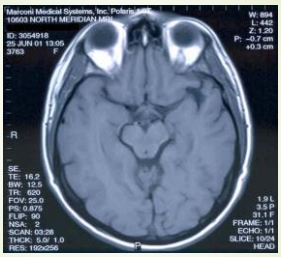
May

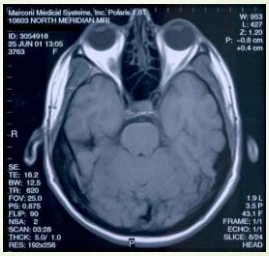
Remember the headaches?

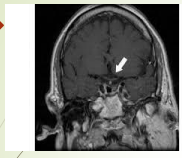
- Referral for neurological consult and imaging
  - Rectangular mass 17X6 mm at the level of the Pons/CN V, left side
  - Termed a "Schwannoma" or a "meningioma."
  - No impingement on optic nerve/chiasm
  - Is there an association with retinal findings?
    - Seemingly unrelated
    - However, literature tends to support a possible association (not necessarily causative)

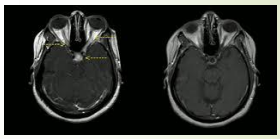
Cerebellopontine Angle











Facts About CPA Tumors

- 6-10% of intracranial tumors
- Vestibular schwannomas and meningiomas two most common types
  - Acoustic neuroma a misnomer
- Arise from Schwann cells
- Connection between vestibular schwannoma and neurofibromatosis type 2 (central).
  - Genetic
  - Connection with development of retinal lesions

Our patient

- Final examination 11 months after the initial presentation and after removal of the tumor
- VA: OD 20/15 OS 20/400
- Screening Visual Field: OS nasal VF loss with partial crossing of midline
  - Patient never returned for threshold testing
  - Patient moved out of area and was lost to follow-up

CNS and Retinitis

- Althaus et al. case report suggests a full neurological evaluation may be indicated with APMPPE cases due to possibility of cerebral vasculitis, which may be manifested later (up to years)
- University of Kentucky study (O'Halloran et al.)
  - 21 males, 10 females ranging in age 8-54, median of 27
  - 35% of patients had antecedent viral illness
  - VAs variable from 20/20 to CF
  - CNS findings ranged from HA to sagittal sinus thrombosis
  - Recommendations: LP/CNS imaging along with Dx of APMPPE
  - Tx: Immunosuppressive agents along with steriods if case is complicated by CNS arteritis

Segway into…

Inflammatory

- Iatrogenic-post-srugical
- Sarcoid
- Ocular toxocariasis
- Tuberculosis
- Histoplasmosis
- Toxoplasmosis
- Syphilis

Tumors

- Choroidal
  - melanoma
    - Amelanotic/melanotic
  - Metastatic
  - Solitary hemangioma
  - Osteoma
- Retinoblastoma

How to tell the difference?

- Case history
- Physical exam
  - Other accompanying characteristic ocular manifestations
- Lab testing/biopsy/imaging

Irvine-Gass

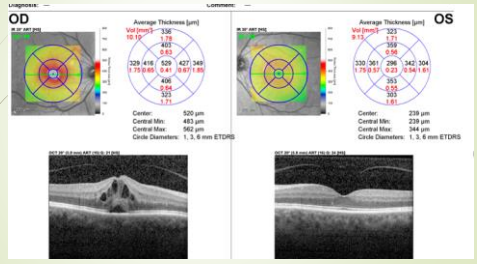
- 72-Year-Old male
  - In Jan of 2016 patient reported for annual exam (established).
    - In 2015 had complained of glare, but tolerable.
    - General health good.
  - CC: "I have problems with glare, especially at night, which has become more noticeable."
    - In addition: "I have been having dizzy spells; they keep changing my BP medication dose, but no answers yet."
  - Noted 2+ NS OU same as previous visit, but patient more symptomatic
    - BAT 2015: 20/30 OD, OS; 2016: 20/100 OD, OS
    - Refer to OMD for cataract evaluation

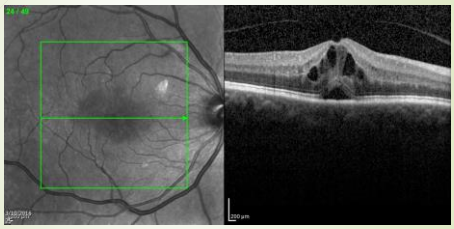
Post-Op 1 week (Feb 2016)

- Patient feels OD vision is worse now than it was before surgery
  - Love hearing that, don't you?
  - VAs 20/25 and 20/25+
    - No problem; must be the healing process
  - Moderate A/C reaction OU
  - Internal health appears normal
    - Continue with steroid as Rx'd; RTC three weeks for one-month post-op or as needed

Post-Op 1 month (March, 2016)

- Patient unhappy with right eye's vision
  - VAs 20/40 and 20/25
  - Macular reflex affected
  - A/C reaction still present (fewer cells, but as much or perhaps an increase of flare)





Cystoid Macular Edema

- Exact cause unknown; most likely inflammatory
- Can be associated with:
  - Retinal vein occlusion
  - Uveitis
  - Diabetes
  - Most commonly seen post-cataract surgery
    - 1-3% of cases
    - 50% chance of second eye involvement if CME in one eye
    - Self-limiting (according to literature)
      - My experience has been some remaining quality of vision deficit even if VA recovery full
  - Treatment varies from topical to injection of steroid or anti-VEGF

Continued

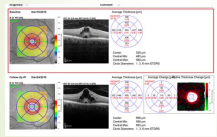
- Increased dose of Durezol
- Included NSAID
- RTC 2 weeks

Two weeks later

- "I have double-vision and my right eye is no better."
  - VA 20/50, 20/30
  - A/C continues to be mildly to moderately inflamed
  - Refer back to surgery center to consider injection given symptoms and OCTs

Two-week follow-up

OD



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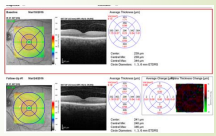
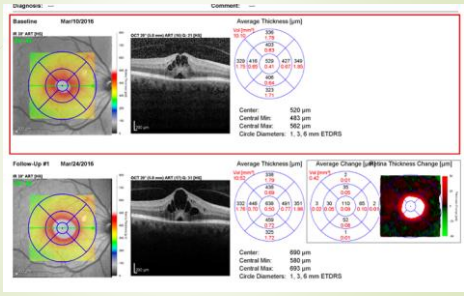
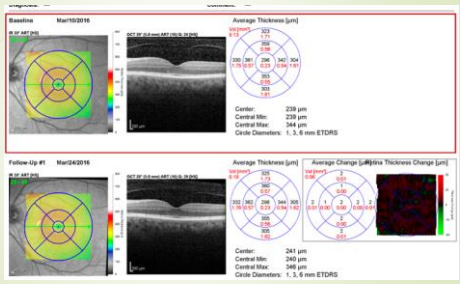
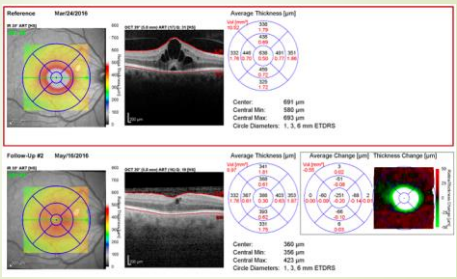


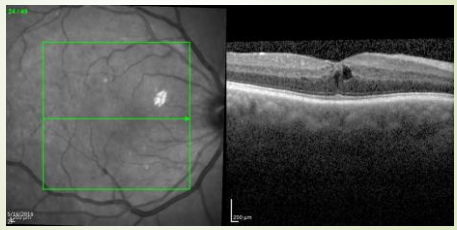




May 2016

- Dr. Kohne's healing touch:
  - Due to improvement, no injection was given, but the treatment was continued using only topical means
  - Patient notices improvement of vision and all the other symptoms
  - VA 20/40, 20/25-



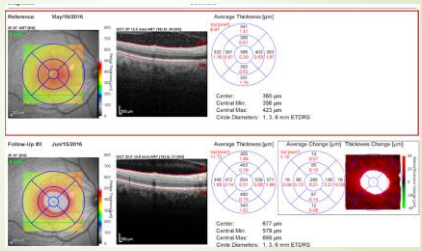


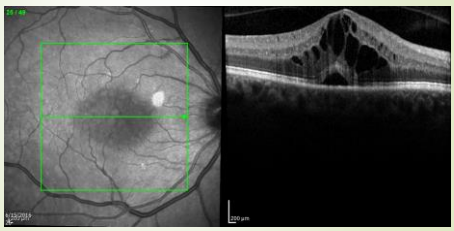
May 2016

- Patient missed a two-week follow-up
  - We called the patient, who stated eye is better
  - Rescheduled visit for June

June 2016

- "My vision is bad again, like it was right after the surgery."
- Compliant with medication, steroid now tapered to only once a day
- VA: 20/40; 20/25
  - Could this be the endpoint of the incident? Could this be as good as we can make it (considering the decrease in CME in May)?
  - We are not done





Continued

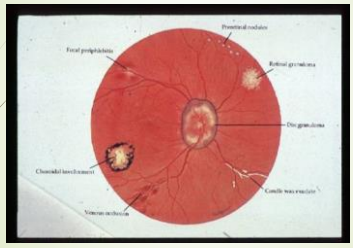
- Called the surgery center immediately and spoke to one of the doctors (the surgeon was not available).
- Requested another evaluation and re-emphasized consideration of injection.
- As of writing this, patient is at the surgery center being prepared for the first injection.

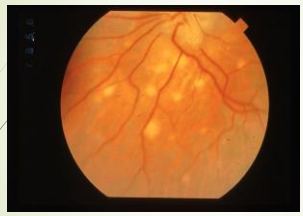
Sarcoidosis

- African American b/w 20-50 y/o in US
- Usually females
- Multifaceted disease
- 27-50% have ocular involvement
- Systemic: Erythema nodosum, parotid involvement, facial palsy, arthritis, liver, spleen, etc.

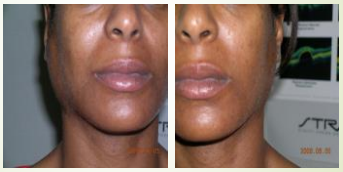
Sarcoid

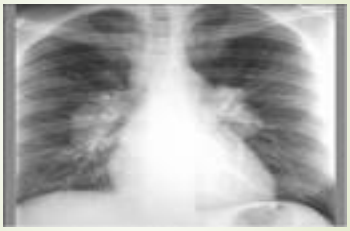
- Ocular manifestations
  - Granulomatous anterior uveitis
  - Conjunctival granuloma
  - Band keratopathy
  - "Candlewax" drippings on veins
  - Choroidal granuloma
  - Periphlebitis
- Laboratory testing
  - ACE, serum lysozyme, chest X-ray revealing hilar adenopathy gallium scan, increased calcium





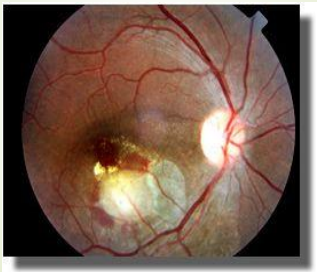
Parotid Gland Enlargement R > L





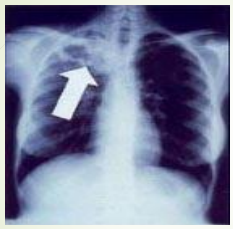
Ocular Toxocariasis

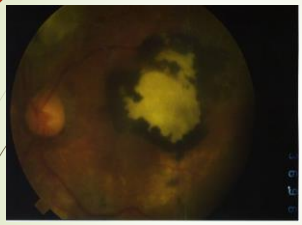
- Parasite – infects dogs and cats
- Occurs between 2-40 y/o, but predominantly between 7-8
- Usually from eating dirt or under-cooked meat
- Found in eye secondary to decreased VA, leukocoria or strabismus
- Mild-severe iritis/vitritis
- Diagnosed by ELISA for Toxocara

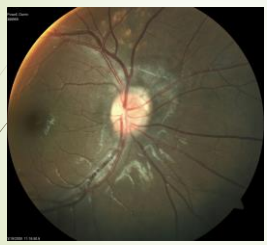


Tuberculosis

- Chronic infection caused by *Tubercle bacilli*
- *Mycobacterium tuberculosis* also a causative agent
- Rare, but can cause uveitis
- Ocular manifestations
  - Yellow-white, multifocal choroidal lesion
  - Granulomatous/non-granulomatous anterior uveitis
- Laboratory testing
  - PPD test, chest X-ray





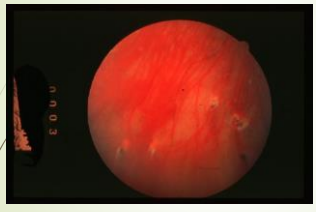


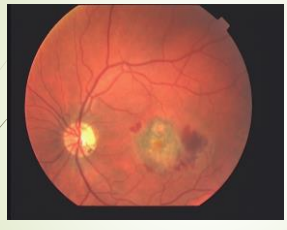
Histoplasmosis

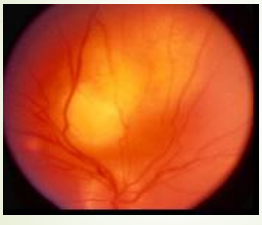
- Fungus that presents in 20-50 y/o whites living near or visited the Ohio-Mississippi River Valley (and several other river valleys throughout the world)
- 60% of cases bilateral
- **lacks any uveitic (anterior and posterior) involvement
- Affected patients over 30 have higher risk for macular involvement
- Predominantly a clinical diagnosis

Histoplasmosis

- Ocular manifestations
  - Classic triad
    - Peripheral punched-out lesions
    - Peri- or circum-papillary atrophy
    - Neovascularization of the macular (sub-retinal net formation)
  - Lacks any uveitic manifestations
- Testing
  - FA if net is present or suspected
  - OCT
  - HLA-B27

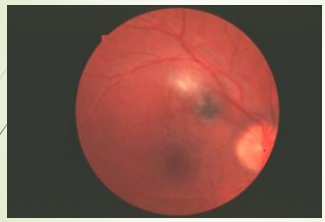






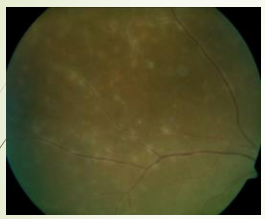
Toxoplasmosis

- Obligate intracellular protozoan
- Most common cause of posterior uveitis
- Usually passed from mother to child
- The mother usually gets the parasite by exposure to feces
- Can be caused by under-cooked meat, breathing in spores or drinking contaminated water
- Ocular manifestations
  - Unilateral, multifocal chorioretinal lesions
  - Severe posterior uveitis response
  - Classic "headlights in the fog"
- Laboratory testing
  - Complimentation fixation test (positive active disease), Sabin-Feldman methylene blue test, ELISA, HIV test



Syphilis

- Ocular manifestations
  - Depends on stage
    - Stage 1: chancre
    - Stage 2: chorioretinitis, uveitis, optic neuritis, conjunctivitis, episcleritis, scleritis
    - Stage 3: optic atrophy, IK, Argyll Robertson pupil
  - Lab testing
    - RPR, VDRL (current infection), FTA-ABS, MHA-TP (past infection)



Retinal and Choroidal Tumors

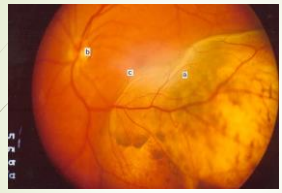
- Usually distinguished by their characteristic appearance
- Ocular testing done such as FA, ICG, OCT, and B-scan, as well as laboratory and systemic testing to rule out markers

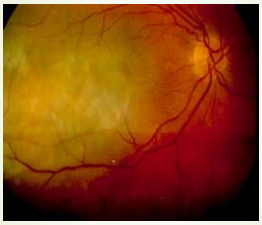
Choroidal Melanoma

- Most common primary intraocular tumor in adults
- Accounts for 80% of all uveal malignant melanomas – mainly due to vascular supply
- Women slightly more at risk
- More often unilateral and diagnosed in people over 50 years of age
- More common in whites

Continued

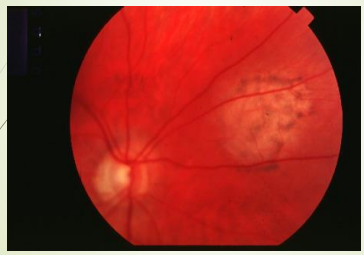
- Typically asymptomatic initially. However, patient may present with one or more of the following:
  - blurred vision, VF loss, and flashes/floaters
- Two varieties
  - Circumscribed
  - Diffuse
- Radiation is the most common treatment modality
- Enucleation is an option for high risk melanomas
  - Concern about seeding must be weighed

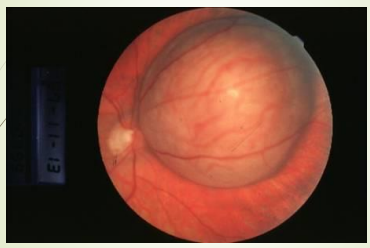


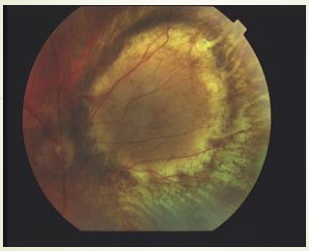


Amelanotic Choroidal Melanoma

- Appearance
  - Indistinct borders
  - Yellowish-white
  - Fairly large diameter and thickness
  - Can produce overlying drusen
  - Brown mottling surrounding the lesion
- Testing
  - Ultrasound, FA (variable), ICG
  - Systemic testing should be done to completely rule out metastasis
    - Colonoscopy, OB/GYN exam, mammogram, MRI of abdomen and pelvis

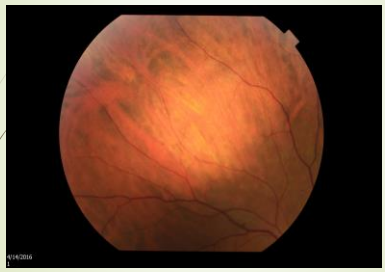


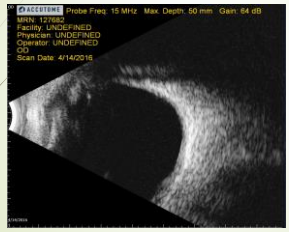




47-Year-Old Woman

- Long-term patient at the clinic
- Long-standing melanotic nevus OS
  - 2 X 1.5 DD, approximately 2 DD superior temporal to macula
    - Stable since first exam in early 2000s.
- Generally healthy, non-smoker, mother of three, no FHx
- April 2016 exam
  - No change in health status
  - No new medications
  - But no one said a thing about OD, including yours truly





Referral

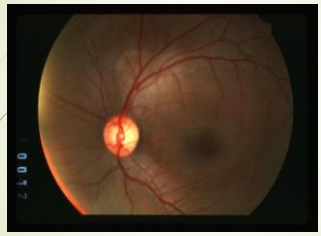
- The lone retinal specialist in town did not accept the patient's insruance
- Referred to OMD, general practitioner
  - Recommended more B-scan
  - Recommended blood work
- Awaiting results and communication from OMD
  - As of July 20, 2016
    - OMD recommends observation only.

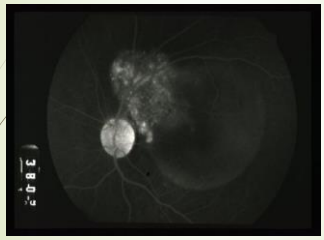
Choroidal Metastasis

- More common than primary malignancies
- Choroid most common site for metastasis (80%), due to high metabolic rate and perfusion, as stated before
- Common primary sites
  - Females: breast
  - Males: lung

Continued

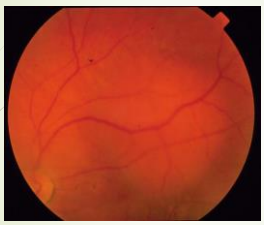
- Ocular manifestations
  - Creamy yellow in appearance
  - Can cause RD
  - Vitreal hemorrhage
  - Can cause proptosis in extreme cases
  - Treatment will depend on size, location, patient age and health
- Tests:
  - Ultrasound, FA (hyperfluorescence in late arterial and early venous)
  - Systemic testing to find primary site if not already known





Solitary Choroidal Hemangioma

- Ocular manifestations
  - Reddish-orange in color with oval or circular shape (classic "tomato catsup" retina
  - Can create subretinal fluid in macula
  - Can cause CME, exudative RD and secondary glaucoma
  - Usually no treatment unless subretinal fluid threatens the macular region
- Testing:
  - B-scan (solid dome tumor, high reflectivity, acoustic solidity)
  - FA (early filling, late leakage)



Choroidal Osteoma

- Slow growing intrachoroidal bone-like tumor
- Usually unilateral (~75%)
- Found in healthy young females
- Typically near the optic nerve or in the posterior pole
- Symptoms depend on location

Continued

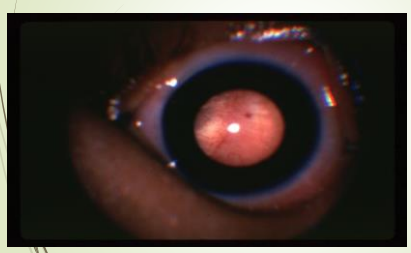
- Ocular manifestations
  - Yellow-white to red-orange in color
  - Edges are scalloped
  - Can cause macular detachment or neovascularization
  - Prognosis depends on location
  - ANY growth suggests malignancy until ruled out
- Tests:
  - B-scan (high reflectivity)
  - CT, X-ray, and FA
    - Mottled early hyperfluorescence that intensifies throughout the angiogram cycle

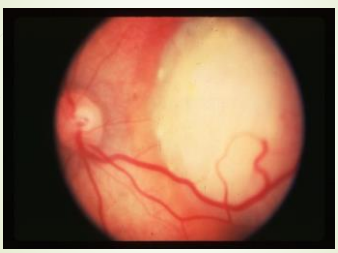
Retinoblastoma

- Most common intraocular malignancy in infants and children
- 2nd most common intraocular tumor in the general population
- Approximately 30% bilateral
- Average age of diagnosis is 18 months
- Can be inherited (only 7%) or sporadic
- Two types:
  - Endophytic (from retina into vitreous)
  - Exophytic (in subretinal space)

Continued

- Ocular manifestations
  - Leukocoria – most common presenting sign
  - Strabismus
  - Dome-shaped lesion
  - Whitish-pink in color
  - Can cause non-rhegmatogenous RD
  - Early detection is crucial due to high risk of metastasis
  - Enucleation is considered as a last resort
- Tests:
  - B-scan (high reflectivity)
  - CT, MRI (to rule out metastasis to brain)





46-Year-Old Man

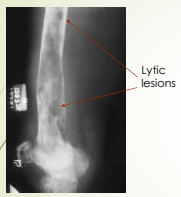
- Another long-standing patient, who presents for annual exam and to have his previously diagnosed retinal hole evaluated (date of initial Dx. 2001 by yours truly).
- Today's exam unusual except on casual observation followed by a question or two, patient informs us that he has developed eczema "for the past 6-8 months."
- All else proceeds normally. We decide to bring the patient back after looking at the nerves and IOPs for a preglaucoma workup simply because it had not been done before.
- Then, we evaluate fundus with BIO for the retinal hole and we see this:

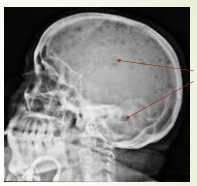
Continued

- I diagnose three pars plana exudative cysts.
  - Associated with multiple myeloma in older African-Americans (our patient is Caucasian and only 46) with symptoms of weight loss (our patient has gained weight), bone and joint pain (patient complains of no aches or pains).
  - However, there is some anecdotal evidence of association with eczema.
- Referred to retinal specialist with recommendation for bloodwork to check markers (increased immonoglobulins).

Multiple Myeloma

- Early symptoms
  - Fatigue and weakness
  - Weight loss
  - Bone pain
- Blood markers:
  - Hypercalcemia
  - Anemia
  - Kidney problems (build-up of toxins)
  - High protein (Immunoglobulin)
- X-rays
  - Weakness spots (lytic lesions)
- CT-Scans, MRIs, PETs





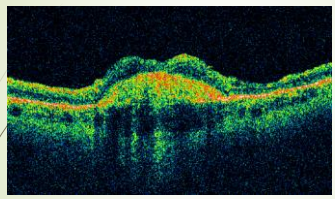
Treatment

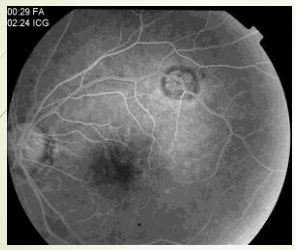
- Most effective is Stem Cell Transplantation (SCT)
  - Recipient cells removed, frozen, and stored
  - Suitable donor cells are a possibility also
  - Existing bone marrow destruction with chemotherapy
  - Replacement with SCs through venous injection
  - Not a cure, but prolongs life expectancy

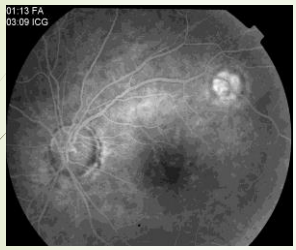
Conclusion

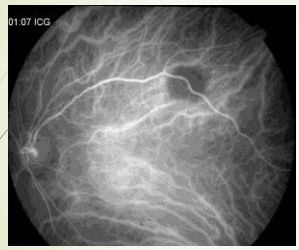
- There is a lot to know. It would be difficult to give the patient a clear-cut answer from one visit, but knowing what the possibilities are makes it easier to discuss the findings. It also makes it easier to discuss the case with the OMD involved.

Whatever happened to our other "routine" exams?



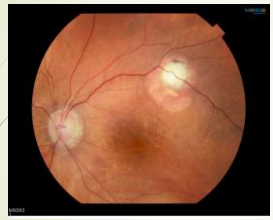


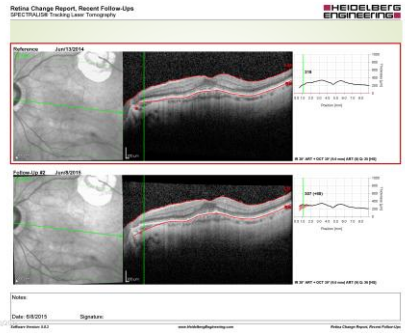




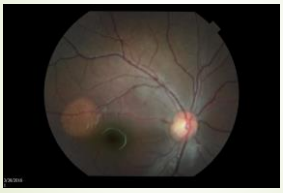
What is SIC Anyway?

- Solitary Idiopathic Choroiditis
  - Yellowish-white lesion of unknown etiology
  - Occurs at any age (usually 20-50)
  - No predilection for gender or age
  - VA usually good unless in juxtapapillary or foveal area
  - IOP and AC normal
  - Patient usually lacks any contributory medical, systemic or ocular history, which makes this a diagnosis of exclusion
  - It contains both active and inactive phases



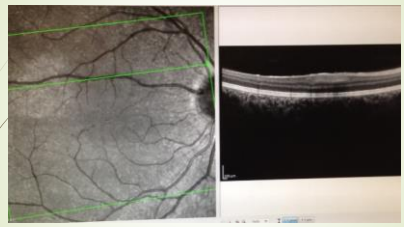


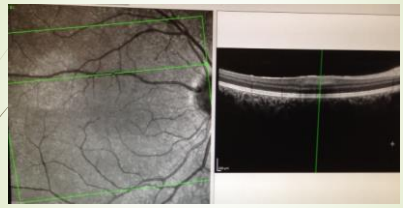
And this one?



13-Year-Old African-American Female

- Previous patient of our pediatric clinic between the ages of 5 and 8
- Last exam, however, 5 years ago
- No other exams any other location according to mother
- *No indication of the "lesion" anywhere in previous encounters*





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