Optometric Education Consultants

Clinical Case Challenge

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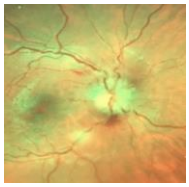
Case 1

- A 27-year-old woman presents urgently complaining of painful vision loss in her right eye.
- She has no known medical history
- She has an edematous optic nerve with hemorrhaging, an afferent pupil defect, superior arcuate scotoma, pain when she moves her eye, and 20/70 visual acuity. Fellow eye is normal



What is the likely diagnosis?

- Demyelinating optic neuritis
- Non-arteritic anterior ischemic optic neuropathy
- Arteritic ischemic optic neuropathy
- Infectious optic neuropathy
- Hereditary optic neuropathy
- Infiltrative optic neuropathy
- Perineuritis
- Papilledema

Case 1

- Infectious optic neuropathy needs to be strongly investigated in a case like this. Neuroretinitis, often associated with cat-scratch disease, will present with a macular star of exudates, but this finding may be missing early in the disease. However, optical coherence tomography (OCT) will show a serous macular detachment early in the disease course.
- In this case, contrast-enhanced MRI of the orbits/chiasm and brain should be ordered to rule out demyelination and neural sheath swelling in perineuritis.
  - Contrast enhanced MRI orbits and chiasm with fat suppression
  - Contrast enhanced brain MRI looking for white matter lesions
- The patient should also be tested for numerous infectious agents including Bartonella, syphilis, Lyme, tuberculosis, herpes, Epstein-Barr, and rickettsioses, to name a few.
  - This can best be done in concert with the patient's primary care physician or an infectious disease specialist.

Diagnosis: Infectious Optic Neuropathy

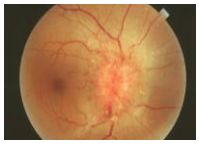
- MRI findings showed optic nerve enhancement possibly consistent with infectious, autoimmune, or granulomatous disease with no evidence of demyelination.
  - "Basically says there is something wrong with the optic nerve"
- Serological testing subsequently revealed very high titers of Epstein Barr Nuclear Antibody IgG and Epstein Barr Capsid Antibody IgG.
- The pathogenesis of infectious optic neuropathies may involve direct involvement of the optic nerve by a pathogen and /or indirect involvement with inflammatory, degenerative, or vascular mechanisms.

INFECTIOUS OPTIC NEUROPATHY

- Syphilis
  - Retrobulbar, papillopathy, neuroretinitis, perineuritis
  - Retrobular, bulbar: severe vision reduction
  - Perineuritis has normal vision, MRI optic sheath enhancement
- Lyme - mimics syphilitic optic neuropathy
  - Bite of mammalian deer tick- can cross react with syphilis
- Toxoplasmosis, HIV/AIDS, CMV
  - Destructive to vision
- Neuroretinitis
  - Typically benign lymphoreticulosis (cat scratch disease)

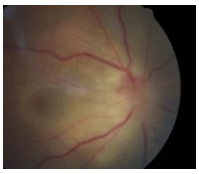
Neuroretinitis

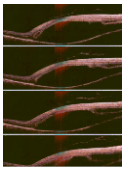
- Mild RAPD compared to vision loss
  - Vision loss more retinal than optic nerve
- Serous macular RD
  - OCT shows subretinal fluid between disc and macula in cases with disc edema only
- Macular star late finding



62 YOF

- 'Strep throat'
- CF @ 8' OD, 20/25 OS – antibiotics x 1 day
- RAPD OD
- Black spot and blurry vision 3 days



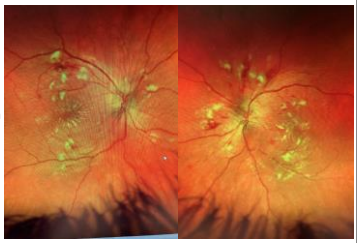


Neuroretinitis/ Infectious Optic Neuropathy

- Many potential etiologies
  - Toxoplasmosis, toxocariasis, measles, syphilis, Lyme disease, herpes simplex and zoster, E-B-V mumps, tuberculosis, malignant hypertension, ischemic optic neuropathy, and leptospirosis, *bartonella* (most common). Fleas are vectors, thus no need for actual scratch.
- Prognosis for visual recovery excellent, especially if the cause is cat scratch disease. Other causes need treatment
  - Most patients will have a return to normal or near normal vision without
  - Antimicrobial therapy may be used to hasten recovery.
    - Rifampin, ciprofloxacin, doxycycline, sulfamethoxazole; doxycycline 100 mg PO BID for one month. Intravitreal injection of Avastin™ (bevacizumab) has been shown to improve both visual acuity as well as decrease macular edema. However, the overall good prognosis of neuroretinitis may not justify this treatment, especially since this information comes from case reports and not controlled clinical trials
  - While antibiotics are frequently used for cat scratch disease neuroretinitis, there are no controlled clinical trials that indicate a better clinical outcome from this therapy. The same can be said for the use of oral steroids and intravitreal anti-angiogenic medications.

Case 2

- 22-year-old female
- First eye exam- blurred vision
- 20/40 OD; 20/70 OS
- No medical history
- Thin build

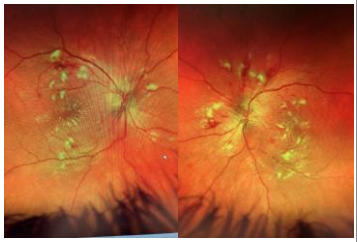


What is the likely diagnosis and plan?

- Bilateral CRVO- coagulopathy workup
- Idiopathic intracranial hypertension- prescribe diamox and weight loss
- Brain tumor- immediate MRI
- Malignant hypertension- BP assessment and ER referral
- Infectious optic neuropathy because that was what the last case was
- Help! I don't know. That's why I'm here

Case 2

- BP 180/144
- Likely diagnosis- malignant hypertension
  - But it *could* be other things



Hypertensive Crises

Hypertensive EMERGENCIES

&

Hypertensive URGENCIES

Hypertensive EMERGENCY

- Severe Hypertension + *End-Organ Damage*
  - Examples of end-organ damage… *hypertensive encephalopathy*, *intracerebral hemorrhage*, *acute myocardial infarction*, *left ventricular failure with pulmonary edema*, *acute coronary syndrome*, *dissecting aortic aneurysm*, or *eclampsia*
- Hypertensive EMERGENCIES require immediate BP reduction (not necessarily to normal ranges) to prevent or limit organ damage.
  - Patients with hypertensive emergencies are often admitted through the ER for aggressive treatment.

Hypertensive URGENCY

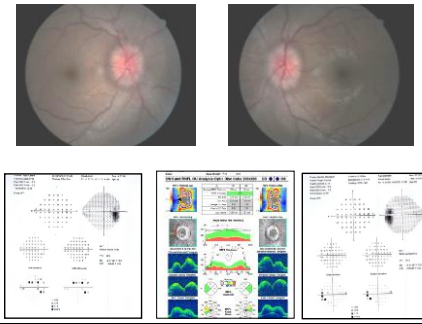
- Severe Hypertension + *NO End-Organ Damage*
  - Typically identified during **routine** evaluation
  - Usually represents chronic hypertension, nonadherence to drug therapy or inadequate treatment by the PCP.

Hypertensive URGENCY

- DOES NOT warrant aggressive BP reduction, as rapid reduction of BP may be associated with significant MORBIDITY:
  - Causes a rightward shift in the pressure/flow autoregulatory curve in critical arterial beds (cerebral, coronary, renal).
  - Can result in marked reduction in perfusion, causing ischemia and infarction; BP must be reduced in a SLOW and CONTROLLED fashion.
  - Patients with hypertensive urgencies are usually treated with oral medications and followed over several days to weeks to evaluate their response to therapy.

Case 3

- 33-year-old-femaleHorizontal diplopia
- Headache
- TVOs 20/day
- Denies OCP, tetracyclines, vitamin A
- Lost 10 lbs- headaches improved
- 118/72
- 5'5"; 160lbs; BMI 26.62



What is the most likely diagnosis?

- Pseudotumor cerebri
- Idiopathic intracranial hypertension (IIH)
- Malignant hypertension
- Intracranial mass lesion
- Venous sinus thrombosis

PTC vs. IIH

- Pseudotumor Cerebri (PTC)
  - Increased intracranial pressure in the absence of an intracranial mass lesion
  - Many causative agents have been identified
- Idiopathic Intracranial Hypertension (IIH)
  - Increased intracranial pressure without an identifiable cause
  - Young, obese females are at risk
- Primary PTC
  - IIH
- Poor CFS drainage

PSEUDOTUMOR CEREBRI DIAGNOSIS

- Si/SX: consistent with increased ICP
- Papilledema
- Normal neurological examination
  - except for cranial nerve 6 abnormalities
- Neuro-imaging: Normal without evidence of hydrocephalus, mass, or structural lesion, thrombosis
- Normal CSF composition

Elevated LP opening pressure
- Adults: > 250 mm CSF
- Children: > 280 mm CSF
> 250 mm CSF if not sedated/obese

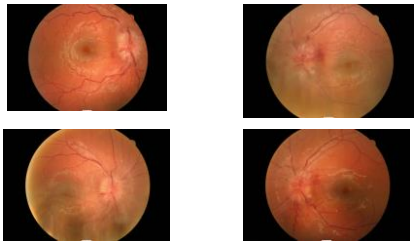
PSEUDOTUMOR CEREBRI DIAGNOSIS

- LP may be deferred if:
- MRI/MRV shows no additional abnormalities and has characteristic findings of flattened globe and empty sella.
- No evidence of fever or acute infection
- Typical profile

Case 4

- 16-year-old-male
- Vision has been fluctuating for 6 weeks
- PCP feels it's normal growth spurt
- Mom feels it's migraines as there is a strong family history
  - Still wants eyes checked
- VA 20/20 OD/OS uncorrected
- Externals: normal
- Meds
  - Inhaler for asthma PRN
  - Minocycline 50 mg BID PO for acne

IIH: "It's not rare if it's in your chair"



Polling question 4: What is the most likely diagnosis?

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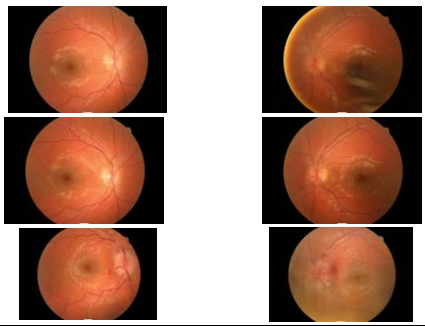
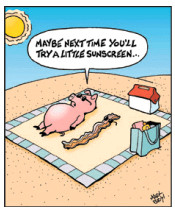
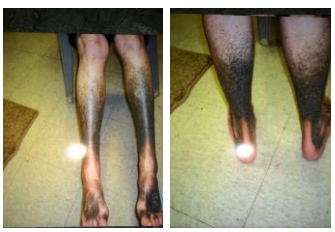


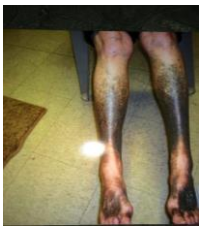
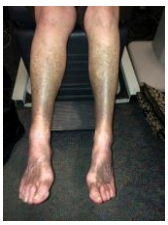
Precautions With Oral Tetracycline Analogs

- Enhanced photosensitivity
- Avoid in children and pregnancy (Category D)
- Can enhance Coumadin
- Can enhance the action of digoxin
- ?Long term use with increase risk of breast cancer?
  - 1 paper/study, not regarded as highly reliable study
  - Further investigation discredited the association
- Benign intracranial hypertension, reported cases
  - 17 cases from 1978-2002

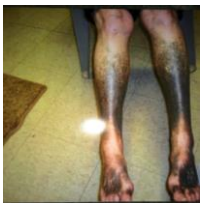
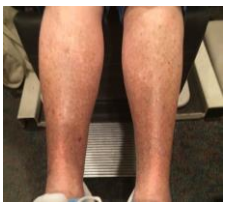




6 Month Later

1 Year Later

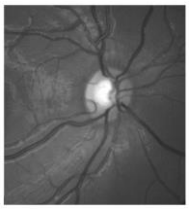
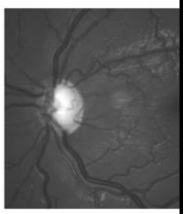



Case 5

- A 13-year-old female was referred for painless reduced vision (20/40) in her left eye with a concurrent abnormal screening visual field, reportedly elevated intraocular pressure (IOP), and an afferent pupillary defect.
- Her previous exam was 3 weeks earlier and she had been previously referred to an ophthalmologist over a year earlier by another optometrist, but her mother did not know why and did not take her.

Case 5

- The key piece of diagnostic information was her IOP
- 28 mm Hg OD and 43 mm Hg OS by Goldmann applanation.
- Pachymetry was slightly thick at 593µ OD and 595µ OS
- There were no biomicroscopic or gonioscopic abnormalities and both angles were open.

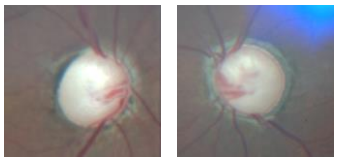
What is the most likely diagnosis?

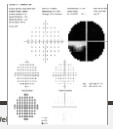
- Orbital tumor OS causing cupping and elevated IOP
- POAG
- Juvenile open angle glaucoma
- Congenital glaucoma

JOAG

- OAG diagnosed during childhood
- Occurring between age 3 years and early adulthood (40 yrs?)
  - Pressure rise occurs after 3rd birthday, but before 16th birthday
- More aggressive course in JOAG than POAG.
- Anterior segment and anterior chamber angle normal
- Appears to be autosomal dominant

Edna, 20/20 OD, OS, Age 37





JOAG

- Developmental immaturity of the trabecular meshwork
  - Endothelial cells lining the inner wall of Schlemm's Canal lack giant vacuoles
  - Thick, compact tissue on the anterior chamber side of Schlemm's canal
  - Abnormal deposition of ground substances.
- Essentially normal appearance
- Note: There are no 'normal tension' JOAG pts
  - IOP *will* be high

Medical Management in Children

- Topical beta blockers: Safe and effective in children.
- Prostaglandin analogs: Safe, well tolerated, but not very effective.
  - Best for older children with JOAG.
- Topical CAI: Safe and effective
  - Probably the best option.
- Brimonidine: Effective, but unacceptable side effects. Should not be used

Understanding Pediatric Glaucoma

Primary Infantile/ congenital

- Abnormal angle
- Globe enlargement
- Corneal edema
- Onset near birth
- Megalocornea
- Symptomatic
  - Blepharospasm, photophobia, lacrimation

Open angle/ secondary

- Normal angle
- Normal axial length
- Clear cornea
- Onset later
- Normal corneal size
- Asymptomatic

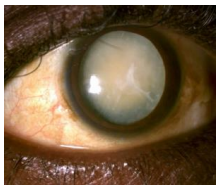
Case 6

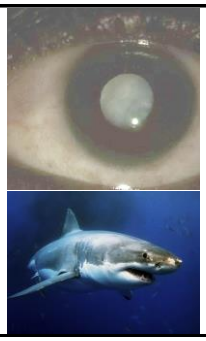
- A 53-year-old man who had been treated for advanced glaucoma presented with slowly progressive, painless vision loss in his right eye. He had missed his visits for the past year, though he had been obtaining medication refills through his pharmacy. His vision in this eye is Light Perception. A year earlier, it was 20/200 and 3 years earlier it was 20/70 with fixation loss from glaucoma.

What is the most likely cause?

- Advancing glaucoma and snuff out of vision
- An orbital tumor
- Angle closure
- Cataract
- Help! I don't know. That's why I'm here

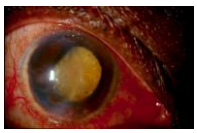
Case 6

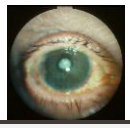


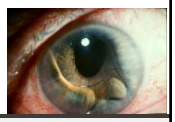


Lens Induced Glaucomas

- Phacolytic
- Phacomorphic
- Lens particle
- Phacoanaphylactic (retained lens fragments)
- Ectopia lentis







Case 7

- A 25-year-old woman was involved in a minor automobile accident where she was hit by another driver. The accident was reportedly minor, with no initial injury to either driver, and both cars were able to be driven away. She felt that she experienced only a mild-to-moderate bump during the accident with no head trauma or loss of consciousness. However, immediately upon waking the next morning, though she had no physical pain, she experienced profound double vision.

What is the likely cause?

- A subarachnoid hemorrhage
- A third nerve palsy
- Orbital fracture
- Fourth nerve palsy
- Sixth nerve palsy
- Help! I don't know. That's why I'm here

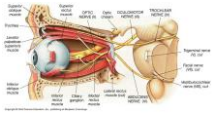
Case 7

- She described the diplopia as vertical and worse at near. She had a distinct right hyper deviation which, on alternate cover test, worsened in left gaze and right head tilt. This was a signature motility of a cranial IV (trochlear) palsy.



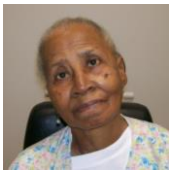
CN IV Anatomy

- Exits the midbrain posteriorly and decussates
- Longest course
- Travels around tentorium, through cavernous sinus, through SOF
- Most prone to trauma



CN IV Palsy

- Longstanding CN IV palsy may present with diplopia from decompensation
  - Observe old photos for head tilt (*Facebook Tomography*)
- Rule of 40-30-20-10

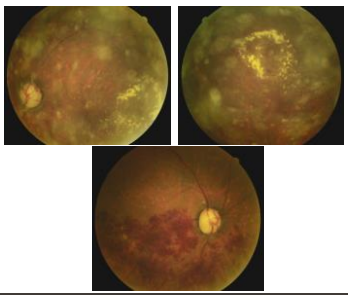


CN IV Management

- Isolated, non-traumatic:
  - Evaluate for ischemic diseases
  - Non-ischemic causes of non-traumatic, isolated CN IV palsy rare
  - Look for longstanding decompensation
    - Increased vertical vergences
    - Old photos

Case 8

- An 84-year-old African-American man with a history of glaucoma presented for consultation and care.
- Review of past records indicated that he had also been diagnosed the previous year with dry age-related macular degeneration (AMD) OD and wet AMD OS with a circinate ring of exudates temporal to the left macula.
  - He reported never following through with a retinal consultation for the AMD.



How many things do you see wrong here?

- One
- Two
- Three
- Four
- Five
- Six
- Seven

Does this patient have wet AMD?

- Yes
- No

Case 8

- Dilated fundus examination revealed an extensive hemi-retinal vein occlusion in the right eye, advanced glaucomatous disc damage OU, scattered retinal drusen OU, asteroid hyalosis OS, disc collaterals OS and curiously a circinate ring of exudates OS nearly identical to the description from the previous record.
- Macular degeneration in a patient of African descent is a very uncommon occurrence.
- The unchanging ring of exudates would not be consistent with wet AMD.
- Careful inspection revealed a saccular dilatation within the circinate exudates consistent with a retinal arterial microaneurysm (RAM).

Case 8

- Retinal arterial macroaneurysms are acquired saccular or fusiform dilatations of the large arterioles of the retina. Patients who develop RAM are typically in the 50–80-year-old age range.
- RAM are caused by a break in the internal elastic lamina of the arteriole wall, through which serum, lipids and blood exude into the surrounding retina. The natural course of RAM typically involves spontaneous sclerosis and thrombosis, particularly after hemorrhaging.
  - Asymptomatic non-leaking RAM may be monitored at 4-6 month intervals.
  - If there is leakage in the form of exudation and/or hemorrhage that does not threaten the macula, then monitoring at 1-3 month intervals is indicated.
  - If hemorrhage threatens or involves the macula or if there is persistent macular edema reducing vision or creating visual field loss, then direct photocoagulation of the RAM may speed resolution. Intravitreal bevacizumab has shown promise as an effective therapy for complicated RAM and cases with submacular exudation.

Case 9

- A 46-year-old woman
- She had a history of breast cancer 5 years earlier and was using tamoxifen.
- She reported that she got some cleaning fluid in her right eye 4 weeks earlier.
  - She said that she had some moderate pain and visual blur which subsided, but then several hours later her vision significantly diminished in this eye. She attributed this to the chemical getting in her eye.
  - She was now 20/400 OD (previously she was 20/20).
  - RAPD OD
  - Pale optic disc with attenuated retinal arterioles
  - OCT showed profound retinal thinning.

What is the likely diagnosis?

- Arteritic anterior ischemic optic neuropathy
- Non-arteritic anterior ischemic optic neuropathy
- Central retinal artery occlusion
- Chemical injury to the optic nerve
- Orbital tumor
- Optic neuritis

Case 9

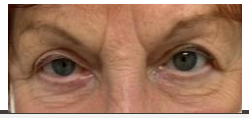
- Based upon her history of sudden, painless vision loss, a pale disc, attenuated retinal vessels, and profound retinal thinning on OCT, she was diagnosed with a central retinal artery occlusion (CRAO). The chemical exposure had no bearing.
- Potential causes:
  - Heart disease, cardiovascular disease, giant cell arteritis, smoking, obesity, carotid artery disease, bacterial endocarditis.
  - Emboli from various sources travel through the vascular system becoming lodged inside the central retinal artery obstructing the flow of blood to distal tissues.
    - Calcific emboli are most likely to cause retinal artery occlusion and are often cardiac in origin.
  - Etiologies related to malfunctioning clotting factors in blood such as antiphospholipid disease, factor VIII abnormality along with protein S and C alteration are also possible etiologies.

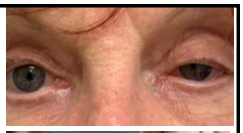
Case 9

- 46-year-old patient, conditions such as giant cell arteritis, atherosclerosis, hypertension and diabetes are not likely
- Tamoxifen is a selective estrogen receptor
- Tamoxifen has been noted to cause a crystalline retinopathy and should be in the differential of possible causes. However, this patient had a unilateral occurrence and no crystalline maculopathy, thus a direct cause is unlikely.
- Tamoxifen associated with thromboembolic events
- CRAO can be caused by inherited or acquired thrombophilia, especially homocysteinemia. Thrombophilia is also caused by exogenous estrogens (oral contraceptives, tamoxifen).
  - Breast cancer create a pro-thrombotic state with an increased risk of thromboembolism.
- Tamoxifen-associated thrombophilia likely precipitated her CRAO
  - As the occlusion was over a month old, urgent referral to a stroke unit stroke was not required. Communication was made with the patient's primary care physician to inform of the retinal vascular event and initiate investigation for a pro-thrombotic state and discuss the possible association with tamoxifen use and prevention of future thrombotic events.

Case 10: 78 YOF

- Sudden onset of ptosis OS
- Immediately following parathyroid surgery
- Headache and eye pain
- Dilation lag and positive Iopidine test





Discussion

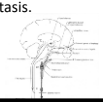
- What is Horner's Syndrome?
  - a triad of clinical signs arising from disruption of sympathetic innervation to the eye and ipsilateral face that causes *miosis*, upper lid *ptosis*, mild elevation of the lower lid, and *anhydrosis* of the facial skin.

What is the most likely cause?

- Lung cancer
- Carotid dissection
- Direct surgical trauma to the nerve
- Migraine
- Help! I don't know. That's why I'm here

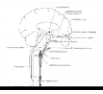
Horner's Syndrome: Etiologies

- First-order neuron disorder: Stroke (e.g., vertebrobasilar artery insufficiency or infarct); tumor; multiple sclerosis (MS), and, rarely, severe osteoarthritis of the neck with bony spurs.
- Second-order neuron disorder: Tumor (e.g., lung carcinoma, metastasis, thyroid adenoma, neurofibroma). Patients with pain in the arm or scapular region should be suspected of having a Pancoast tumor. In children, consider neuroblastoma, lymphoma, or metastasis.



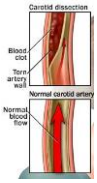
Horner's Syndrome: Etiologies

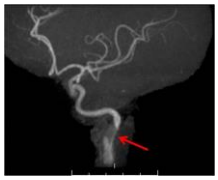
- Third-order neuron disorder: Headache syndrome (e.g., cluster, migraine, Raeder paratrigeminal syndrome), internal carotid dissection, herpes zoster virus, otitis media, Tolosa–Hunt syndrome, neck trauma/tumor/inflammation, prolactinoma.
- Congenital Horner syndrome: Trauma (e.g., during delivery).
  - **Facebook tomography**
- Other rare causes: Cervical paraganglioma, ectopic cervical thymus



Carotid Dissection

- *A 3rd-order Horner's and ipsilateral head, eye, or neck pain of acute onset should be considered diagnostic of internal carotid dissection unless proven otherwise.*





Carotid Dissection

- Carotid artery dissection presents with the sudden or gradual onset of ipsilateral neck or hemicranial pain, including eye or face pain
- Often associated with other neurologic findings including an ipsilateral Horner's syndrome, TIA, stroke, anterior ischemic optic neuropathy, subarachnoid hemorrhage, or lower cranial nerve palsies
  - 52% with ocular or hemispheric stroke with 6 days
    - 67% within first week; 89% within 2 weeks; none after 31 days
- Horner's from suspected carotid dissection should go to ER

Case 11: 72 YOWF

- New onset flashing lights OD (OU?)
  - No floaters
- 20/20 OD, OS
- PERRL (-) RAPD
- CF Full OD, OS
- MF IOL OU
- Asthma; hyperlipidemia

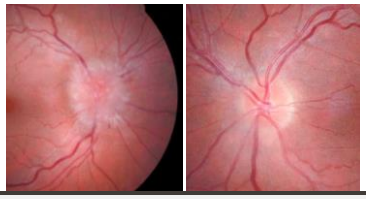
What is the most likely etiology

- Retinal detachment
- PVD
- Migraine
- Some other weird thing because this is too easy
- I don't know and that's why I'm here

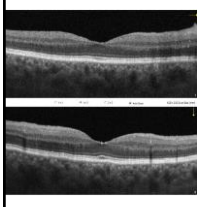
Case 11: 72 YOWF

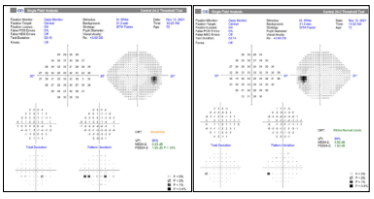
- New onset flashing lights OD (OU?)
  - No floaters
- 20/20 OD, OS
- PERRL (-) RAPD
- CF Full OD, OS
- MF IOL OU
- Asthma; hyperlipidemia

Hint: she has been to a Lyme endemic area



72 YOWF





What is the most likely etiology?

- Compressive tumor
- Infectious optic neuropathy
- Papilledema
- Ischemic optic neuropathy/ GCA
- Infiltrative optic neuropathy
- Some other weird thing
- I don't know and that's why I'm here

Now it gets complicated

- Presumptive diagnosis: Infectious optic perineuritis
- MRI Brain & MRI orbits and chiasm w contrast and fat suppression
- Normal ☹
- Ask for neuroradiology re-read specifically looking for ONH sheath enlargement and perineuritis (*It's gotta be Lyme*).
- Normal again☹☹
- Now on to serology (working with PCP)
- CBC, ESR, CRP, ANA, RPR, FTA-Abs, ACE, zoster IgM and IgG, rubella IgG and IgM, toxoplasmosis, Lyme, Bartonella henselae and quintana panels were all normal. The only abnormalities were herpes simplex IgG and IgM, and Epstein Barr IgG and IgM. Subsequent EBV ab VCA IgM was normal
  - Old infections of HSV and EBV- non-contributory