

Optic Nerve Grand Rounds: "You've Got Some Nerve!"

Joseph Sowka, OD, FAAO,
Diplomate



1



DISCLOSURE

Dr. Joseph Sowka is a member of the advisory boards for Novartis, Glaukos, Allergan, and B&L. Dr. Sowka has no direct financial interest in any of the diseases, products or instrumentation mentioned in this presentation. He is a co-owner of Optometric Education Consultants. www.optometricedu.com

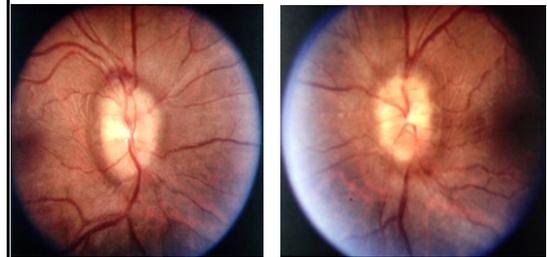


2

28 YOF

- Presents with intermittent blurred vision & visual "grey-outs", intermittent horizontal diplopia, and chronic headache steadily worsening X 2 weeks
- MHx: "white coat hypertension", shoulder injury X 6 mos
- Meds: Flexeril® 10 mg BID PRN
- Height / weight: 5'3", 220 lbs.
- VA: OD 20/20, OS 20/20
- Pupils & motility: normal

3



What questions do you want to ask?
What tests do you want to perform?

4

POLLING QUESTION 1

5

28 YOF

- Additional hx: Dull "ringing" in ears
- BP: 142/100
- SLE: unremarkable
- T_A: OU 16 mm Hg
- VF: blind spot enlargement & nasal step defect OU
- Serology Normal
- Imaging: small ventricles, otherwise normal
- LP: O.P. = 510 mm H₂O; all CSF studies normal
- DX: Pseudotumor cerebri (PTC)

6

DEFINITION:



PAPILLEDEMA: EDEMA OF THE OPTIC DISC, SPECIFICALLY RESULTING FROM ELEVATED INTRACRANIAL PRESSURE.

7

PAPILLEDEMA: SIGNS & SYMPTOMS

Signs:

- bilateral disc edema
 - superior & inferior aspects of discs affected FIRST
 - obliteration of optic cup
 - hemorrhages common
 - absence of SVP
 - Paton's folds
- highly variable VF defects
 - enlarged blind spot (early)
 - arcuate defects and constricted (late)
- NO RAPD typically
- VA near normal

Symptoms:

- Visual:
 - transient visual obscurations
 - intermittent horizontal diplopia
- General:
 - headache common
 - nausea & vomiting
 - dizziness
 - tinnitus

8

PAPILLEDEMA TYPES:

- **Acute**
 - Hemorrhages, exudates, hyperemia, RNFL edema
- **Chronic**
 - Minimal hemorrhage/exudate. Collateral vessels may be present
- **Atrophic**
 - Eventually occurs if papilledema remains chronic. Optic disc pallor



9

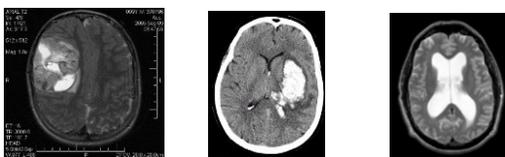
PAPILLEDEMA PATHOPHYSIOLOGY

- **Disc edema results from axoplasmic stasis**
 - intracellular fluids, metabolic by-products accumulate and are regurgitated at the level of the optic nerve head
 - in papilledema, cerebral edema is effectively transmitted along the common meningeal sheaths of the brain and optic nerve producing an engorged, swollen disc.

10

PAPILLEDEMA PATHOPHYSIOLOGY

- **Associated with intracranial abnormalities:**
 - increased brain volume (intracranial mass lesion)
 - increased intracranial blood volume
 - Intracranial hemorrhage
 - increased CSF volume
 - Hydrocephalus
 - Ventricular blockage by mass lesion



11

PAPILLEDEMA MANAGEMENT

- **Rule out "swollen disc masqueraders"**
 - ultrasonography can be invaluable in differentiating ONHD
 - also consider color, margins, SVP, vasculature, etc.
- **Acute papilledema constitutes a medical emergency**
 - Immediate neuro-imaging to rule out an intracranial mass.
 - if imaging is normal, lumbar puncture to measure CSF pressure and exclude meningitis or other disease processes is necessary.
- **Atrophic papilledema with significant vision/field loss:**
 - urgent measures must be undertaken to prevent blindness
- **Papilledema accompanied by any neurologic abnormalities, fever or stiff neck:**
 - Possible serious underlying neurologic abnormality, intracranial infection or bleed requiring immediate medical attention.

12

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 CSF drainage**

13

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

14

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**

15

PSEUDOTUMOR CEREBRI MANAGEMENT

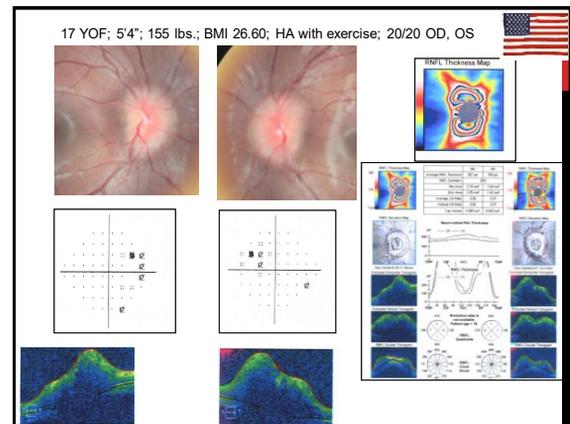
- **No visual loss**
 - Symptomatic headache therapy
 - Acetazolamide 500 mg tid
 - Weight reduction
- **Mild visual loss**
 - Acetazolamide 500 mg tid
 - Furosemide, Topiramate, Zonisamide
 - Weight reduction

16

PSEUDOTUMOR CEREBRI MANAGEMENT

- **No/ Mild visual loss**
 - Prognosis
 - Excellent (all signs and symptoms, visual loss)
 - 6-9 months
 - Follow-up and visual fields
- **Role of weight loss**
 - Treat the primary problem
 - 10% weight loss
 - Prevent recurrence
 - Keep the weight down

17

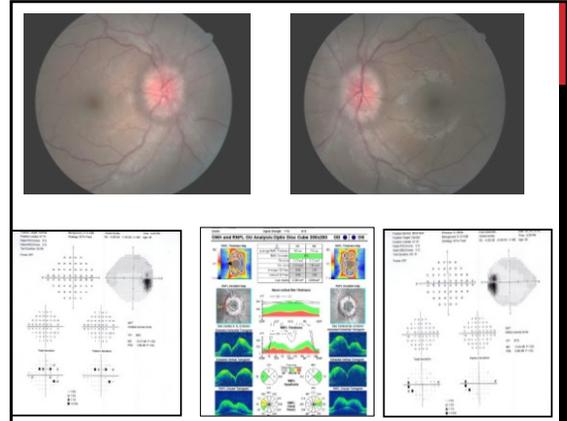


18

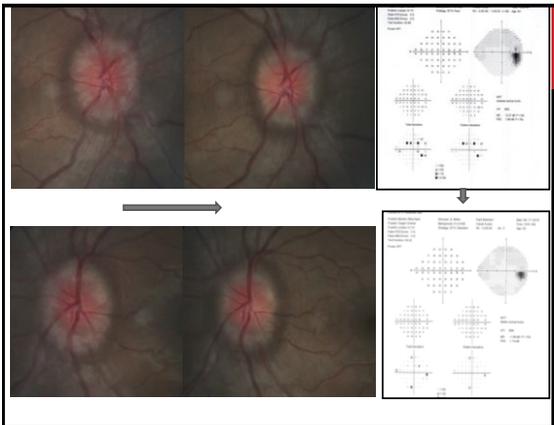
33 YOF

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

19



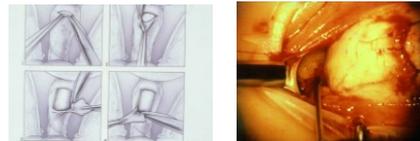
20



21

PSEUDOTUMOR CEREBRI MANAGEMENT

- **Severe, or progression of visual loss**
 - Optic nerve sheath decompression (ONSD)
 - High-dose IV steroids and acetazolamide
 - Lumboperitoneal shunt
 - Failed ONSD
 - Declined ONSD
 - Intractable headache



22

POLLING QUESTION 2

23

REMEMBER

- Not all elevated discs are swollen, not all swollen discs are edematous, and not all edematous discs are papilledema
- True papilledema is a medical urgency and should be treated as such with a search for the cause.
- Many conditions can present with papilledema, including intracranial mass lesion, hydrocephalus, VST, PTC
- PTC is a diagnosis of EXCLUSION.



24

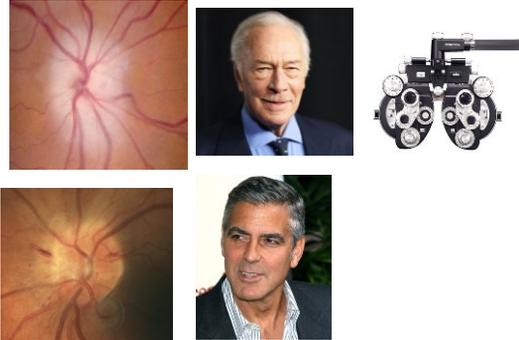
ODE TO A SWOLLEN DISC

When you think the disc is swollen,
The vessels north and south will appear stolen.
Not all elevated nerves are edematous,
Just like not all snakes are venomous.
Your thoughts should go to papilledema,
But infection and inflammation should still be in your schema.
MRI, MRV and LP,
are soon to be.
Remember, pseudotumor is a diagnosis of exclusion,
Female and firm does not make pseudotumor a forgone conclusion.
Brain tumors can exist when the pseudotumor profile is classic.
Do the evaluation so they don't end in a casket.

Joseph Sowka, OD

25

WHICH IS BETTER? ONE OR TWO?



26

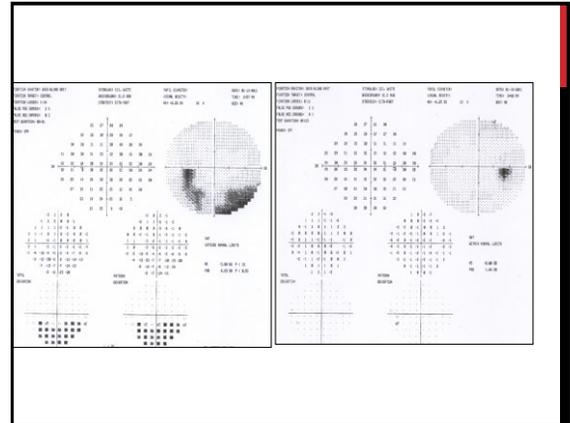
48 YOM

Painless loss of visual field OS

- 20/20 OD, OS
- Noticed upon waking

Med Hx: Unremarkable, except for viral illness 3 weeks before

27



28



29

74 YOM

• Presents with 'worst headache of his life'

- Sees: PA, ED physician; cardiologist; NP;
 - *3-week period*
- Histories: Eye ache; jaw pain, scalp pain, facial pain, somnolence; malaise; jaw claudication
- Diagnoses: TMJ; Lyme disease
- "vasculitis such as temporal arteritis highly unlikely", "Not GCA"
 - However, ESR and CRP ordered and elevated- never reviewed
- Ultimately OD makes diagnosis
- End result?

30

POLLING QUESTION 3

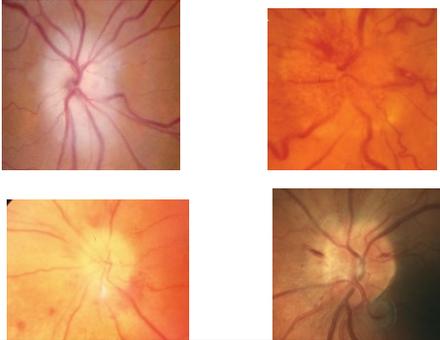
31

ANTERIOR ISCHEMIC OPTIC NEUROPATHY

- Hypoperfusion of the posterior ciliary arterial supply to the anterior optic nerve head.
- May be arteritic (AAION) or non-arteritic (NAAION)
- Mechanical factors and atherosclerotic disease play a role in the non-arteritic form while vasculitis contributes in the arteritic form.
- Unilateral presentation but high incidence of subsequent contralateral involvement
 - AAION

32

AAION VS NAAION



33

NAAION

- Risk factors:
 - Hypertension, diabetes, atherosclerotic disease, small optic nerves
- Inferior field defects
- Hyperemic swollen nerve- disc at risk
- Progressive moderate vision loss with potential recovery
- Late 30s/ early 40s and beyond
- Painless



34

AAION

- Pallid optic nerve swelling with flame hemorrhages, arteriole attenuation and NFL infarcts
- Pain (of some sort)
- Severe optic nerve dysfunction
- Visual field defects
- Giant cell arteritis/ PMR- risk factors
- Typically 70s, uncommon under 60
- High risk bilateral involvement



35

DIAGNOSIS

- Careful history: Must directly ask about nonvisual symptoms
 - Headache (present in over 90%), scalp tenderness, jaw claudication (almost diagnostic), ear pain, arthralgias, temple pain and/or tenderness, malaise, intermittent fevers
- Examination
- Laboratory studies
 - Erythrocyte sedimentation rate
 - Lowered by statins and NSAIDS
 - C-reactive protein
 - Not affected by statins and NSAIDS
 - Elevated platelet count

36

AAION

DIAGNOSIS

Prodrome, GCA symptoms
Elevated ESR/CRP (combination of the two gives high specificity [97%])
Elevated platelet count (acute phase reactant)
Ophthalmoscopy
Fluorescein angiography
Temporal artery Biopsy

TREATMENT

Prompt steroids and hydration
Consider IV when vision loss present

- Very effective in prevention of second eye
- Occasionally restores vision

37

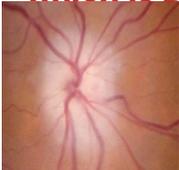
AAION VERSUS NAAION

- Think AAION >> NAAION
- Systemic symptoms of GCA
- TVOs/amaurosis
- Elevated
 - ESR/CRP
 - Platelets
- AION + cilioretinal artery occlusion
- Evidence of posterior ciliary artery occlusion on FA
- Early massive vision loss
- Chalky white optic disc edema

Hayreh SS, Podhajsky PA, Zimmerman B. Occult giant cell arteritis: ocular manifestations. Am J Ophthalmol. 1998;Apr;125(4):521-6.

38

WHICH IS BETTER? ONE OR TWO?



Bilaterally blind



Residual field loss, but otherwise not bothered

39

ODE TO AN ISCHEMIC NERVE

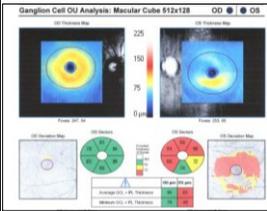
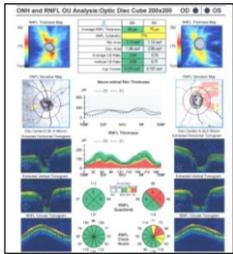
When your patient's optic nerve is ischemic
 You better hope the disc is hyperemic.
 In Non-arteritic no treatment is needed
 And life will rarely be impeded.
 But if the disc is swollen and pale,
 And vision is an epic fail
 If the patient is sixties, seventies or eighties
 You will feel heat like in Hades
 ESR and CRP are required
 And steroids must be acquired
 Remember, when you see a choked disc
 Always assess the giant cell risk

Joseph Sowka, OD

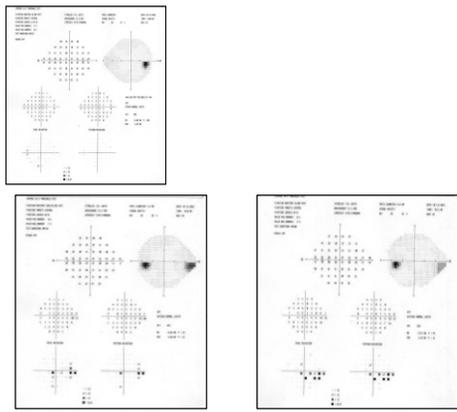
40

29 YOF

- Referred for glaucoma evaluation due to suspicious cupping- no complaints
- IOP 12 mm OD, 13 mm OS

41

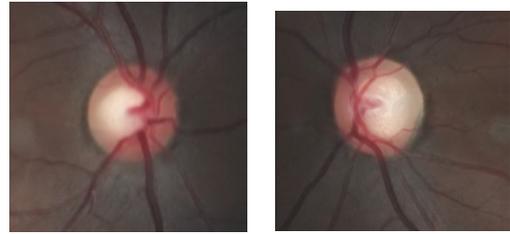


42

29 YOF

- *"Now let's get serious"*
- 20/15 OD, OS
- IOP: 12 mm Hg OD, 13 mm OS
- CCT: 493 OD, 488 OS
- Gonio normal OU
- +RAPD OS

43



Segmental disc pallor OS

44

POLLING QUESTION 4

45

OPTIC ATROPHY

- **Primary optic atrophy**
 - Uniform nerve fiber degeneration, resulting in glial replacement but no architectural alteration of the optic nerve head.
 - Disc appears chalky white, but the margins remain distinct and retinal vessels appear normal.
 - Trauma and compression (e.g. tumor) causes
- **Secondary optic atrophy**
 - Results from pathological chronic disc edema
 - malignant hypertension, papilledema, or infiltrative diseases like leukemia or sarcoidosis.

46

OPTIC ATROPHY

- **Consecutive optic atrophy**
 - Degenerative retinal conditions
 - Retinitis pigmentosa, pathological myopia and central retinal artery occlusion.
 - Pale, waxy disc, normal margins and marked attenuation of the arterioles.
- **Temporal disc pallor**
 - Toxic/ nutritional (bilateral) or demyelinating optic neuropathy (optic neuritis)

47

OPTIC ATROPHY

- **Numerous potential etiologies**
 - Infarction, infection, infiltration, inflammation, trauma, toxicity, metabolic dysfunction or direct compression of the nerve or chiasm
- **Evaluation:**
 - MRI studies should be obtained of the orbits, the optic chiasm and the brain with and without contrast, fat suppression for orbits, in a high field scanning unit.
 - Contrast dye (gadolinium) is beneficial in discerning malignant lesions, demyelinating plaques indicative of multiple sclerosis.

All cases of optic nerve pallor/ optic atrophy must be investigated or explained

48

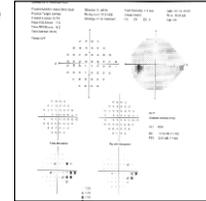
OPTIC ATROPHY

- **Systemic causes of optic atrophy**
 - sarcoidosis, tuberculosis, Behçet's disease, lymphoma, leukemia, systemic lupus erythematosus, nutritional or metabolic disorder (e.g. pernicious anemia, folate deficiency), syphilis, Lyme disease, and antiphospholipid antibody syndrome.
- **CBC, ESR, ACE, ANA, serum cardiolipin, serum homocysteine, serum B12 and folate levels, and rapid plasma regain (RPR) for syphilis.**
 - Additionally, chest x-rays could prove helpful in suspected cases of TB or sarcoidosis.

49

29 YOF- OUTCOME

- **MRI orbits- normal (limited/ poor study)**
- **Repeat MRI brain- no lesions**
- **Lupus panel, ANA, DS DNA, ESR, metabolic panel, Vit B12/folate- normal**
- **RPR, HIV- non-reactive**

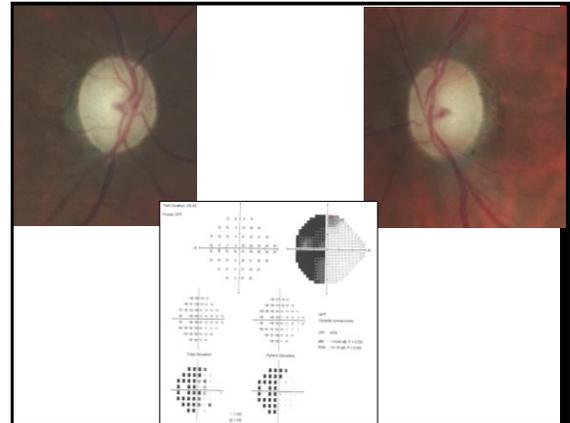


50

A FAMILY AFFAIR

- **56 YOBF**
- **Dx POAG OU 5 years ago**
- **Slowly progressive vision loss**
- **LP OD; 20/30 OS**
- **Used combo med- ran out months ago**
- **IOP: 19 mm OD, 18 mm OS**
- **CCT: 560; 544**

51



52

TECHNIQUE: MRI of the orbits without and with gadolinium contrast.

Magnetic resonance imaging was performed on the brain utilizing sagittal, axial and coronal T1-weighted images. Subsequently, axial T2-weighted dual-echo images were acquired. Sequences were obtained without and with gadolinium contrast.

FINDINGS: The globes are normal in signal and morphology. The optic nerves have a normal MRI appearance. The extra-ocular muscles are normal in signal and morphology. The intraconal and extraconal fat planes are preserved. The visualized portions of the paranasal sinuses are well aerated.

There is a cystic and solid enhancing mass within the sella with dimensions of 36.06 mm in craniocaudal dimension and anterior posterior dimension of approximately 26.3 mm. The mass is partially cystic with septations but has solid enhancing components and elevates the optic chiasm.

IMPRESSION:

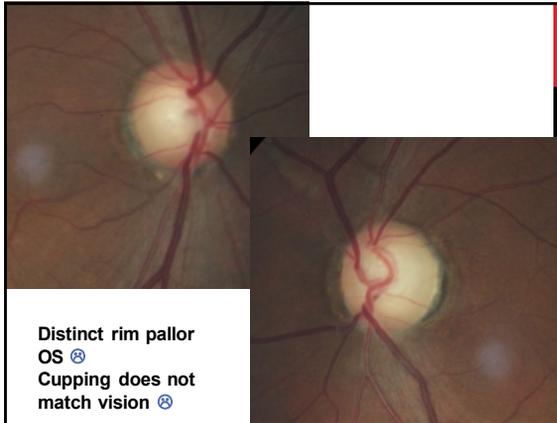
ENHANCING CYSTIC SEPTATED MASS WITHIN THE ADENOHYPHYSIS OF THE PITUITARY WITH DIMENSIONS OF 36.06 MM IN CRANIOCAUDAL DIMENSION AND ANTERIOR POSTERIOR DIMENSION OF APPROXIMATELY 26.3 MM. DIAGNOSTIC CONSIDERATIONS INCLUDE PRIMARILY FOR MACROADENOMA BUT ADDITIONAL DIAGNOSTIC CONSIDERATIONS INCLUDING CRANIOPHARYNGIOMA CANNOT BE EXCLUDED. THERE IS ALSO INCIDENTALLY MILDLY DILATED BILATERAL LATERAL VENTRICLES.

53

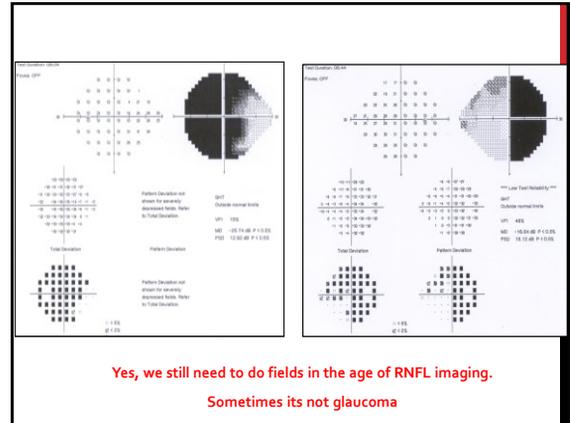
54 YOM

- **Referred for glaucoma management**
- **Told he had glaucoma 6 years earlier - no Tx**
- **20/30 OD; HM OS**
- **30 mm Hg OD; 23 mm Hg OS**

54



55



56

COMPRESSIVE OPTIC NEUROPATHY

- Results from compression of the optic nerve within orbit, at the orbital apex, chiasm-secondary to:
 - Space occupying orbital lesions, including tumor masses
 - Infiltrated extraocular muscles (Graves' ophthalmopathy) in thyroid disease (most common)
- Unilateral with orbital masses, can be bilateral in Graves' disease
- Presents with slowly progressive, variable vision loss; variable proptosis and motility restriction

57

COMPRESSIVE OPTIC NEUROPATHY

- Optic nerve may be initially hyperemic with retinal edema, tortuous vessels, and associated hemorrhages; with prolonged compression, may see pallor and optic disc collateral vessels
- Visual fields consistent with papilledema in early stages, ischemic optic neuropathy/ glaucoma in later stages
- Increased concentric 'cupping' can occur
 - Compression causes pallor; glaucoma causes notching
- Management involves orbital imaging and serum thyroid profile if Graves' suspected

58

ODE TO A CUPPED DISC

Oh, to have a cupped disc pink.
That my friend hath a glaucomatous stink.
But to have a cupped disc pale,
Call this glaucoma and you shall fail.
Disc and field damage that is one-sided
Simply cannot be abided.
It might be trauma, infarct or meningioma.
But if the rim is cut always remember,
Nothing notches a nerve like glaucoma

Joseph Sowka, OD

59

42 YOF

- Sudden painless loss of vision OS x 1 week
 - getting worse, not getting better
 - began as dimming, then rapidly dropped off
- BVA: 20/20 OD; 20/400 OS
- PERRL (+) RAPD OS (mild)
- Conf. Fields: FTFC peripherally OD, OS
- Amsler: Central/cecocentral scotoma OS
- SLE: normal OU
- IOP: 18 mm Hg OD, 19 mm Hg OS

60



61

42 YOF

- No known HIV risk
- Recent illness: Severe flu with malaise, fever, and lymphadenopathy 4 weeks antecedent.
- No tick bites or rashes
- Exposure to cats
- Serology:
 - FTA-ABS/RPR; HIV, Lyme, toxoplasmosis, toxocariasis, PPD: Negative
 - *Bartonella henselae* titers: positive
- Dx: Cat scratch neuroretinitis

62

INFECTIOUS OPTIC NEUROPATHY

- Syphilis
 - Retrobulbar, papillopathy, neuroretinitis, perineuritis
 - Retrobulbar, 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)

63

NEURORETINITIS

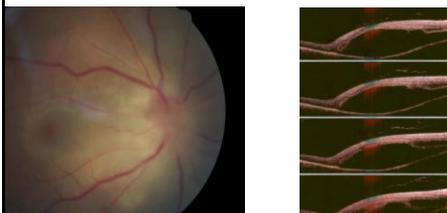
- Mild RAPD compared to vision loss
 - Vision loss more retinal than optic nerve
- Serous macular RD
 - OCT shows subretinal fluid between disc and macula in cases with disc edema only
- Macular star late finding



64

62 YOF

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



65

NEURORETINITIS

- Many potential etiologies
 - Toxoplasmosis, toxocariasis, measles, syphilis, Lyme disease, herpes simplex and zoster, 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.
 - 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.

66

ODE TO AN INFECTED NERVE

When the vision is poor and the APD mild,
It's often the bite of something wild.
If the disc is swollen and macular swelling
great,
Its neuroretinitis and the star comes late.
Syphilis and Lyme are alike,
and can cause similar titres to spike.
One is transmitted sexually and the other not,
Unless the patient is weirder than you thought.

Joseph Sowka, OD

67

THANK YOU FOR YOUR ATTENTION.
ALWAYS REMEMBER TO RECYCLE AND
PROTECT THE PLANET THAT WE WILL
ULTIMATELY LEAVE TO KEITH RICHARDS



77