Conundrum: Is This Really Glaucoma?

IS THIS REALLY GLAUCOMA? WHY DOES IT MATTER?

- Treating a disease that they don’t have
  - Expense and adverse effects
- Treating one disease when its really another
  - Vision loss and potentially worse
- Not treating a disease that they do have
  - Vision loss
- In reality, when encountering patients with mimicking and confounding conditions, the diagnosis is challenging

RULE #1

- Pallor in excess of cupping indicates something other than glaucoma
- When the rim tissue is pale, suspect some cause other than, or in addition to, glaucoma.
- Rim pallor in glaucoma is very rare and is the exception, not the rule.
  - Disc pallor can only be accepted as part of glaucoma when other potential causes have been eliminated

NOT ALL ‘OMAS’ ARE GLAUCOMA

- Pituitary adenoma
- Craniopharyngioma
- Meningioma
- Glioma
- Ischemioma
  - Anterior ischemic optic neuropathy (AAION - cup enlargement but devastating vision loss and disc pallor)
- Retinaloma
  - Retinal infarcts
- Congenitaloma
- Coincidentaloma
- Misdiagnosoma
**RULE #2**

- Nothing notches a nerve like glaucoma.
- Focal damage to the neuroretinal rim is very specific to glaucoma.
  - Tumors don’t notch a nerve, nor do inflammations, infections, ischemia, etc.

**RULE #3**

- In glaucoma, the field should match the nerve
  - The field is allowed to be better than the nerve, but not worse.
  - Look for something else:
    - Look for neurogenicity in field loss pattern
    - Don’t forget the retina

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**CASE**

- 72 YOM
- Long term glaucoma suspect OS
- Fields and imaging not changing
- Treated intermittently for NTG
  - IOP ranges from 09 – 12 OU
- Problem: Nerve does not match field
  - Mild disc pallor

Violates Rules 1-3

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**CASE: 29 YOM**

- Treated for glaucoma for 4 years based upon disc appearance and field loss
- Hx of uveitis OD
  - Reports that his vision was reduced during episode “very foggy”
BY THE WAY, HE ALSO HAD THIS OD...

CASE: 29 YOM

- Normal disc, RNFL, GCC
- Chorioretinal scar and hx consistent with toxoplasmosis
- PVD
- Field loss due to retinaloma
- Stopped meds- 18 mm OD, 16 mm OS; CCT 570 OD; 557 OS
- Retinaloma/ Misdiagnosoma
Age younger than 50 years was 93% specific for nonglaucomatous cupping.

Eyes with nonglaucomatous cupping associated with intracranial masses had significantly lower levels of visual acuity than did patients with glaucoma.

- Visual acuity less than 20/40 was 77% specific for nonglaucomatous cupping.

Eyes with glaucoma had significantly less neuroretinal rim pallor, larger CDR, greater vertical elongation of the optic cup, and greater frequency of peripapillary atrophy and disc hemorrhage than eyes with nonglaucomatous cupping.

- Disc hemorrhage was 100% specific for glaucoma

Pallor of the optic nerve in excess of cupping is a highly specific sign of nonglaucomatous cupping (90.4%)

"THE CUPPED DISC: WHO NEEDS NEUROIMAGING?"

Patients with mass lesions:
- Visual acuity less than 20/40
- Vertically aligned visual fields defects
- Optic disc pallor in excess of cupping
- Age younger than 50 years

CASE: 56 YOF

- 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

DIAGNOSING GLAUCOMA IN ANOMALOUS DISCS

- Rules of disc analysis rarely work in anomalous discs
- Look at other clues and risk factors
- Recognize that we will likely make diagnostic errors
- Ensure that errors have least detrimental effect to patient

72 YOM

- Amblyopia OD and bilateral dense cataracts
- Undergoes cataract surgery OS
- 20/50 PO OS- no complications
- Pt very unhappy with outcome
- Diagnosed with advanced NTG
  - Peak IOP 22 mm; CCT: 536, 531
  - Referred to glaucoma specialist- treated to low teens IOP

72 YOM: CHALLENGES

- Doesn’t believe he has NTG
- Semi-retired attorney who has:
  - Time
  - Money
  - Excellent internet access
  - Will stop at nothing to discover what is wrong.

MARKED TILTED DISC SYNDROME OU
CONGENITAL-OMA

CASE 7: 72 YOM

- Evaluation including neuroimaging unremarkable
  - Macular OCT normal
- Travels country consulting
  - Conflicting opinions
- Significant field loss OU
- Possibly glaucoma as well as congenitaloma (Double-oma)
  - Continue treatment- “Do No Harm”
**JP: 38 YOF**
- Referred for glaucoma eval in 2002 after failing LASIK screening
- Had been treated since mid 20s for glaucoma
- IOP in mid-upper teens off meds
- CCT: 459 OD; 469 OS
- Anomalous nerves with mild field loss

**JP: NOW 49 YOF**
- Congenitally anomalous nerves with field loss
- Monitored for 11+ years
- Field changes late
- Pt now treated with IOP 09 mm OD; 10 mm OS
- Pt had/had congenitaloma and now has glaucoma
  - Doubloma

**SIMILAR...YET DIFFERENT**
- 45 YOF
- Referred for glaucoma evaluation
- IOP never exceeds mid-teens
- CCT: 554 OU
- Marginal effect of meds

**CONUNDRUMS**
- Field loss due to anomaly, glaucoma, or both?
- Progressive or congenital?
- Mid-teen IOP and poor medical response
- Treatment or observation?
**CONCLUSIONS**

- Disc pallor indicates something other than glaucoma
- Rim obliteration/ notching indicates glaucoma
- Glaucoma without risk factors is suspicious
- Differentiating glaucoma from non-glaucoma is challenging when risk factors (IOP) are present
- Fields and nerve should match
- If discs are anomalous, mistakes can be made. Err on the side of caution

**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

**ANSWER:**

- Things have to make sense. If the imaging findings do not fit with the anatomic and functional correlates of pathophysiologic change, trust your own knowledge and judgment.
- When in doubt, repeat the imaging study and the visual field or both.

**Conundrum:**
The diagnostic imaging doesn’t agree with my diagnosis? Now what?

**OCT TO VERIFY GLAUCOMA – THE OPTIC NERVE HEAD?**

Using OCT to Verify Early Glaucoma

A healthy, 70-year-old African American man was referred for evaluation for pigment dispersion. The patient had a moderately elevated cup-to-disc ratio at 0.75, as per his optometrist. His IOP was 15 mm Hg OD and 14 mm Hg OS. This patient was a glaucoma suspect, so I wanted to get good baseline data. His visual field and central visual field tests were normal, but his OCT scan was abnormal.

To verify the OCT, I carefully examined his optic nerves and found that his cup-to-disc ratio was 0.85 x 0.85 OS and 0.85 x 0.80 OS.

**RED DISEASE – A NEW CLINICAL NON-ENTITY**

A supratentorial, non-glaucomatous masquerade disease

Afflicts the educated patient (especially with Internet access) with good health care plans and/or wealth

Debilitating to the patient and painful for the visual care provider to treat

2006. Journal of Irreproducible Results and Senseless Studies
WHAT DO YOU MAKE OF THESE...?

Garbage in, Garbage out.

HELP! THE DIAGNOSTIC IMAGING DOESN'T AGREE WITH MY DIAGNOSIS!
- Low risk OHTN
- Local OD wants imaging for baseline

OCT RNFL NORMAL...

...but markedly abnormal
GCC OS

Same patient, same day, same quality, GCC now normal

Signal strength: 10/10 OD, OS on both images

CASE: 62 YOHM

- Asymptomatic; 20/20 OD; OS
- TA 30 mm OD, 28 mm OS
  - Isolated measurement
  - 12-17 mm OD, 13-17 mm OS
    - 11 visits
- Gonio: open OU w/o abnormalities
- CCT: 597 OU

Don’t make clinical decisions based upon bad data
So, What are your thoughts?

Debate: Treat or Observe?
Debate: Why the disparate findings?
Debate: Why the isolated IOP elevation?

GREEN DISEASE - AN INSIDIOUS CLINICAL ENTITY

A glaucomatous process masquerading as non-disease Afflicts inexperienced, poorly-educated, and lazy doctors who simply want a machine to make all clinical decisions for them Debilitating to the patient and painful for the visual care provider, but a boon for malpractice attorneys

2015. Journal of Irreproducible Results and Senseless Studies

HELP! THE DIAGNOSTIC IMAGING DOESN'T AGREE WITH MY DIAGNOSIS!

• 56 YOM - Glaucoma suspect since 2012
Is this person really a glaucoma 'suspect'?
A example of Green Disease

GREEN DISEASE
RED + GREEN = YELLOW DISEASE?

PANOMAP VERSION 8.0
OCT IMAGING TAKE HOME POINTS

- Serial overlays/imaging to determine baseline (intra-session) noise
- Good signal strength
- Good segmentation without errors
- Optic nerve head exam for disc hemorrhage, pallor, myopic, and tilted nerve heads
- Determine structure-function correlation
- Follow all ancillary tests visual fields and optic nerve head photos for progression

CAUTIONS ABOUT IMAGING

- No current technology is better than the human eye and common sense
- Beware of “Red Disease”
- Treat Real Disease and not Red Disease
- Don’t miss green disease
- Know the limitations of the technology: normative database, reproducibility, resolution, quality of imaging
- Technologies come and go

Conundrum: WHEN IS SURGERY WRONG FOR THE PATIENT?

- When the risk of surgery is greater than its expected benefit.
- When it is more dangerous to undergo a surgical procedure than to continue on the same medical treatment.
- When you would not recommend the same intervention to your family members

GLAUCOMA SURGICAL DECISION MAKING

- Establishing the course of treatment
  - Is the disc or field status stable or worse?
  - If progression has occurred, over what time period?
  - What is the rate of change?
  - What is the risk of visual disability in the patient’s lifetime?
  - Is the patient aware of either decreased central visual acuity or peripheral visual field loss?
    - Classic question: Is it the cataract or the glaucoma or the age related macular degeneration?

IMPORTANT QUESTIONS ABOUT VALUE OF SURGICAL INTERVENTION

- How far to go?
  - Does the patient value the visual acuity of Hand Motions or Light Perception or remaining visual field?
  - What is the status of the fellow eye?
  - Is glaucoma a primary condition or related to a cause (proliferative diabetic retinopathy, central retinal vein occlusion, trauma)?
  - Has a family member become visually disabled from glaucoma?
  - Has a family member lost vision after glaucoma surgery?
**IS FILTERING SURGERY A PANACEA?**

- Trabeculectomy will give low IOP
  - Single digits
- Long history of success
- Technically straightforward process
- Eye never looks/feels the same
- Potential complications

**RISKS OF GLAUCOMA SURGERY**

- **Trabeculectomy**
  - Immediate postoperative period
    - Hypotony – flat anterior chamber, acute cataract, angle closure, choroidal effusion
    - “Wipe out” or “snuff out” syndrome – acute loss of central acuity without obvious intraoperative complication
    - Decreased visual acuity – Patient only knows that they see much worse after surgery
- **Glaucoma drainage implant surgery**
  - Muscle imbalance – noncommittant diplopia

**ADDITIONAL RISKS OF GLAUCOMA SURGERY**

- **Late postoperative period**
  - Posterior synechiae formation – poor dilation
  - Cataract formation
  - Bleb scarring and return of high IOP
- **Very late postoperative period**
  - Endophthalmitis and blebitis
  - Remember “RSVP”
    - R – Redness
    - S – Sensitivity to light
    - V – Vision Change
    - P – Pain

**JUNIOR: 20/60 OD; 20/400 OS 56 YO**

**Edna**

20/20 OD, OS
Age 37

10-2 SS OS
10-2 SS OD
JUNIOR

- Surgery is likely wrong for Edna and Junior
- Risk of wipeout of remaining vision/fixation very real even if low
  - “Doctor, I can’t see your face anymore”
- Medical therapy safest but maybe not surest
  - IOP in mid-to-low teens for both

HAZEL AND JOSEPH

- 87 YOF; 95 YOM - managed for 16 years
- Hazel: 20/20 OD; 20/30 OS; MMT; s/p SLT
  - IOP: 17 mm OD; 20 mm OS
  - CCT: 472 OD, 474 OS
- Joseph: 20/25 OD, OS
  - Cosopt, xalatan, and alphagan
  - IOP 11 mm OD, 13 mm OS
  - CCT 473 OD, 473 OS

HAZEL

JOSEPH
JOSEPH

HAZEL AND JOSEPH
- For whom is surgery right and for whom is surgery wrong?

SUMMARY
- Pts with bare fixation are at high risk of surgical morbidity
  - At some point even aggressive surgeons will decline
    - “Better God than I take their vision”
- Surgery is wrong for your patient when someone you trust as your surgical consultant would not recommend the same procedure to their own family member.

Conundrum:
Help! My patient has a disc hemorrhage and the pressure is 12 mm. Now what?

DISC HEMORRHAGES
- Likely mechanical
- Resolves within 6 weeks. This is the reason that the incidence is difficult to determine.
- Can be recurrent and, if it recurs, it typically is in the same place on the disc each time
- Disc hemorrhages do not constitute a diagnosis of glaucoma nor a progression or conversion to glaucoma or an endpoint for any major glaucoma

Not all hemorrhages of the disc are disc hemorrhages.
RISK FACTORS: DISC HEMORRHAGES

Inferior, inferior temporal, superior, and superior temporal regions of the disc are most susceptible and account for virtually all true glaucomatous disc hemorrhages.

Typically occurs where notches and RNFL defects occur. Hemorrhages at other areas of the disc (nasal and temporal) tend to not be associated with glaucoma.

OTHER CAUSES OF ‘DISC’ HEMORRHAGES

- PVD
- HTN
- Anemia
- Diabetes
- Vascular occlusion
- Subarachnoid bleed
  - Terson’s syndrome
    - Subretinal and intraretinal
    - May be juxtapapillary

55 YOF

- Referred for NTG evaluation based upon disc hemorrhage OS
- IOP: 14 mm OD, 15 mm OS
- 0.3/0.3 OU without pallor, notching or RNFL defect
- OCT/ GDx- normal OU

55 YOF

- Disc hemorrhages not characteristic of glaucomatous hemorrhages
- Structure normal
- Plan: observation
These photos are 18 months apart.

Not all hemorrhages of the disc are disc hemorrhages. Make sure that the glaucomatous characteristics are there.

EARLY MANIFEST GLAUCOMA TRIAL
- Disc hemorrhages- predictive of progression
- Treatment was unrelated to the presence or frequency of disc hemorrhages.
  - Disc hemorrhages were equally common in both the treated and untreated groups of patients.
  - Disc hemorrhages don’t occur in all glaucoma pts.
- Disc hemorrhages cannot be considered an indication of insufficient IOP-lowering treatment,
  - Glaucoma progression in eyes with disc hemorrhages cannot be totally halted by IOP reduction.

OCULAR HYPERTENSION TREATMENT STUDY
- The occurrence of a disc hemorrhage increased the risk of developing POAG 6-fold in a univariate analysis and 3.7-fold in a multivariate analysis that included baseline factors predictive of POAG
- Occurrence of an optic disc hemorrhage was associated with an increased risk of developing a POAG end point in participants in the OHTS
  - However, most eyes (86.7%) in which a disc hemorrhage developed have not experienced a POAG end point to date (5 years)

- The incidence of ODH was 0.5% per year during an average of 13 years before the development of POAG and 1.2% per year during an average of 6 years after the development of POAG
- ODH increased the risk of developing POAG 2.6-fold in multivariate analysis
- Randomization to the observation group, older age, thinner central corneal thickness, larger vertical cup-to-disc ratio, higher intraocular pressure, and self-reported black race were identified as risk factors for ODH.

Disc hemorrhages are a significant risk factor for progression, but do not constitute a diagnosis or actual progression. Further, you cannot stop disc hemorrhages from occurring by lowering IOP.
55 YOM

- 2012 presents without complaints
- BCVA 6/6 OD, OS
- IOP:
  - OD: 27 mm; 30 mm
  - OS: 15 mm; 15 mm
- CCT: 536; 531

Treatment initiated
- IOP drops to mid teens OU
- Optic disc change OS noted 4/14
- Therapy amplified
- 7/15: latanoprost and dorzolamide/timolol FC OU
- IOP: 10 mm OU
- CCT: 536; 531

SO WHAT DO I DO WHEN I SEE A DISC HEMORRHAGE?

- (Treated) IOP high teens:
  - Progression documented- increase therapy
  - Risk of visual disability- increase therapy
  - None of the above: increase therapy or monitor for progression then increase therapy
- (Treated) IOP low teens
  - Monitor for progression (if safe)- no change
  - Progression documented or risk visual disability
    - Therapy increase
    - Equal risk of blindness from disease or treatment
THANK YOU FOR YOUR ATTENTION.
ALWAYS REMEMBER TO RECYCLE AND PROTECT
THE PLANET THAT WE WILL ULTIMATELY LEAVE
TO KEITH RICHARDS