THE NECESSITY OF THE NEURO-RETINAL RIM

Presented by Kelly A. Malloy, OD
June 24, 2017

THE OPTIC NERVE EVALUATION

- HISTORY
- VISUAL ACUITY
- VISUAL FIELD DEFECT
- COLOR VISION
- CONTRAST SENSITIVITY
- AFFERENT PUPILLARY DEFECT
  - DISC APPEARANCE
- INTRAOCULAR PRESSURE

OPTIC DISC FEATURES

- NEURO-RETINAL RIM
  - COLOR
  - CUP
  - VESSELS
  - DISC MARGINS

STRUCTURE and FUNCTION

LEFT VISUAL FIELD

TEMPORAL FIELD

nasal
macular

proximal

distal

LEFT DISC

NASAL DISC

Distal

Proximal

Nasal fibers

Macular fibers

Normal NRR Size /
Thickness

- Disc area
  - I > S > N > T
- Elevation

- NEURO RETINAL RIM / OPTIC DISC ASSESSMENT
  - NEURO RETINAL RIM LOSS
  - NEURO RETINAL RIM PALLOR
  - NEURO RETINAL RIM EDEMA
NEURO RETINAL RIM LOSS

CAUSES OF CUPPING

- GLAUCOMA
- NON-GLAUCOMA
  - ISCHEMIA
    - NAION (10%)
    - AAION (50%)
  - COMPRESSION (orbital, sellar)(6%)
  - INFLAMMATION
  - TRAUMA
  - HEREDITARY / MITOCHONDRIAL

What Differentiates Glaucomatous From Non-Glaucomatous Cupping?

DISC APPEARANCE

(Trube et al 1980)

"OMA"
- PALE rim (94% specific)

GLAUCOMA
- PINK rim (87% specific)
- Polar notching
- Rim obliteration
- Rim sharpening
- Peripapillary halo

IS CUPPING ALWAYS A SIGN OF GLAUCOMA?
IS CUPPING ALWAYS A SIGN OF GLAUCOMA?

Non-glaucomatous cupping can present WITHOUT pallor!!

 Especially true with sellar lesions.

Normal Tension Glaucoma

• Diagnosis of Exclusion
• Many things can mimic NTG
  • If IOP has never been elevated, need to rule out other causes
  • Especially if there is optic disc pallor and the VF does not match the ONH, additional work-up is needed
    — Lab testing to r/o infectious, inflammatory, nutritional etiologies
    — Neuro-imaging to r/o mass, abnormal enhancement
    — Carotid ultrasound to r/o stenosis
    — If progressive vision loss, lumbar puncture may be warranted

Possible Labs for Optic Neuropathy

• CBC with differential and platelet count
• C-reactive protein (inflammation)
• ESR (inflammation)
• Lyme titer (if + get Western blot IgG and IgM)
• ANA with reflex titer (auto-immune disease)
• ACE (sarcoid)
• RPR (syphilis) - If (+) RPR, LP done to confirm neuro-syphilis
• FTA-ABS (syphilis)
• Vitamin B 12 (nutritional)
• Folic acid (nutritional)
• Methylmalonic acid (nutritional)
• Homocysteine (nutritional / inflammatory)
• SPEP (Serum Protein Electrophoresis) (possible malignancy)

GLAUCOMA  versus  “-OMA”  

Non-Glaucomatous Optic Neuropathy

• Normal VA
• Normal
• APD less likely
• disc / field DO match
• Rim defect
• Reduced VA
• Reduced color vision
• APD is likely
• disc / field NOT match
• Rim pallor
NEURO RETINAL RIM PALLOR

- Patterns
- Laterality
- Associated Findings
- Causes
- Work-Up

Development of Pallor

Takes about 1 month to develop

Initial Presentation

26 days later

Neuro-Retinal Rim Pallor

- Pathologic NRR = PALE
- Degeneration / atrophy of axons
- Pallor can ONLY occur in lesions from the Lateral Geniculate Nucleus (LGN) FORWARD
- Ganglion cell axons synapse in LGN
- Lesions posterior to LGN cannot cause pallor

CAUSE OF OPTIC ATROPHY?

- DISC MARGINS  (sharp or blurred?)
- COLOR & NERVE FIBER LAYER  (patterns of pallor?)
- VESSELS  (shunts, peripapillary arterioles, attenuation, embolus?)

WHAT CAUSED THE OPTIC ATROPHY?

WHAT CAUSED THE OPTIC ATROPHY?
ASCENDING OPTIC ATROPHY
(Disease Within the Eye)
- PAPILLITIS
- CHRONIC ATROPHIC PAPILLEDEMA
- PERIARTERITIS
  - CRAO
  - AION

DESCENDING OPTIC ATROPHY
(Disease Behind the Eye)
- RETROBULBAR DISEASE
- OPTIC NEURITIS
- TOXIC
- TRAUMA
- COMPRESSION

WHAT CAUSED THE OPTIC ATROPHY?

PATTERNS OF DISC PALLOR
- DIFFUSE
- SEGMENTAL
  - Temporal
  - Wedge shaped
  - Altitudinal
    - Band / Bow-tie (optic tract lesion)

“TATTLETALE DISC PALLOR”
(Quigley, Miller, Green 1983)
GLAUCOMA
ISCHEMIA
COMPRESSION
TOXIC/COMPRESSION

LEFT VISUAL FIELD
TEMPORAL FIELD
nasal
macular
proximal
distal

LEFT DISC
NASAL DISC
Nasal fibers
Macular fibers
CAUSE OF OPTIC ATROPHY?

- **DISC MARGINS**
  (sharp or blurred?)
- **COLOR & NERVE FIBER LAYER**
  (patterns of pallor?)
- **VESSELS**
  (shunts, peripapillary arterioles, attenuation?)

Optochoroidal Shunt Vessels

- Pre-existing channels
- Dilate in response to chronic obstruction of CRV
- Shunt venous flow to choroidal circulation

- Can be congenital (single) – RARE – only 5% (work-up)
- Acquired (usually multiple) in 95% - to edge of disc

Acquired Optochoroidal Shunts

- CHRONIC PAPILLEDEMA
- OPTIC NERVE MENINGIOMA / GLIOMAS, SPHENOID WING MENINGIOMA
- CRVO (nonischemic)
- ADVANCED GLAUCOMA

Patterns of Pallor and Nerve Fiber Layer Loss

- **PAPILLOMACULAR BUNDLES**
  - TOXIC / NUTRITIONAL
  - HEREDITO/DEGENERATIVE
  - DEMYELINATING
  - COMPRESSIVE
- **ARCULATE BUNDLES**
  - glaucoma
  - ischemia
- **NASAL BUNDLES**
  - CHIASM AND RETROCHIASM

Optochoroidal (optociliary) shunts

- LARGE VEINS CONNECTING THE CHOROIDAL AND RETINAL CIRCULATION AT THE OPTIC NERVE HEAD
ACQUIRED OPTOCHOROIDAL SHUNTS

• If the patient has no history or clinical findings of CHRONIC PAPILLEDEMA, CRVO, or GLAUCOMA, then we must rule-out a:
  – MENINGIOMA of the optic nerve sheath or sphenoid wing
    (neuro-imaging indicated)

OPTIC DISC FEATURES

• NEURO-RETINAL RIM
  • CUP
  • COLOR
  • VESSELS
  • DISC MARGINS

• NEURO RETINAL RIM / OPTIC DISC ASSESSMENT
  – NEURO RETINAL RIM LOSS
  – NEURO RETINAL RIM PALLOR
  – NEURO RETINAL RIM EDEMA

DISC DILEMMA

• IS THIS DISC EDEMA?

PSEUDOPAPILLEDEMA

• Hypoplastic optic discs
• Optic disc drusen
• Other types of anomalous optic discs
  – Usually most elevated nasally

OPTIC NERVE HYPOPLASIA

• PALE, PINK, or BLURRED
• “DOUBLE-RING” SIGN
• VESSELS LOOK TOO LARGE
• TILTED / ELEVATED
• SEGMENTAL or PROFOUND
OPTIC NERVE HYPOPLASIA
(Skarf & Hoyt 1984)

- IF BILATERAL IN CHILD
  - 46% DEVELOPMENTAL DELAY
  - DECREASED GROWTH HORMONE
  - HYPOTHYROIDISM
    - NEED ENDOCRINE WORK-UP
    - POSSIBLE NEUROLOGIC WORK-UP

OPTIC DISC DRUSEN

DISC DRUSEN vs. PAPILLEDEMA
or
DISC DRUSEN and PAPILLEDEMA

DISC DRUSEN

- MARGINS: Scalloped, Blurred
- NRR: Central elevation, Lumpy / bumpy
- NFL: Nasal light reflexes
- CUP: Small/ no cup
- VESSELS: Central origin, Trifurcations

A/B scan with 30 degree test

- To confirm subtle edema
- To differentiate papilledema from pseudopapilledema
- To prove need for more invasive work-up, especially in an asymptomatic patient

A/B scan with A and B

- BOTH pt A and B
- Young Women above ideal body weight
- Only symptom is occasional headaches
- (-) SVP
PATIENT B
Results of A/B SCAN with 30 degree test

<table>
<thead>
<tr>
<th>primary gaze</th>
<th>30 degrees lateral</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right Eye:</td>
<td>2.5 mm</td>
</tr>
<tr>
<td></td>
<td>2.5 mm</td>
</tr>
<tr>
<td>Left Eye:</td>
<td>2.5 mm</td>
</tr>
<tr>
<td></td>
<td>2.5 mm</td>
</tr>
</tbody>
</table>

PATIENT A
Results of A/B SCAN with 30 degree test

<table>
<thead>
<tr>
<th>primary gaze</th>
<th>30 degrees lateral</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right Eye:</td>
<td>4.8 mm</td>
</tr>
<tr>
<td></td>
<td>3.1 mm</td>
</tr>
<tr>
<td>Left Eye:</td>
<td>5.5 mm</td>
</tr>
<tr>
<td></td>
<td>2.7 mm</td>
</tr>
</tbody>
</table>

PAPILLEDEMA

MARGINS
- Blurred (ISNT rule)

NRR
- Edema extends out into NFL

NFL
- Temp Paton’s lines

CUP
- Present

VESSELS
- Obscured at margin, no SVP

ACUTE PAPILLEDEMA

CHRONIC PAPILLEDEMA
Can Progress to Chronic Atrophic Papilledema

Features of edema
- Axoplasmic stasis in pre-laminar optic nerve
- Obscuration of retinal vessels coursing over the disc margin
- Paton’s lines temporally
- Extruded axoplasm (in chronic papilledema)
- Can have hemorrhages
Pattern of Edema

- Corresponds with NFL thickness
- Superior, Inferior > Nasal > Temp
- Superior and Inferior NFL swell first
- Last to swell is Temporal NFL

Modified Frisén Papilledema Scale

<table>
<thead>
<tr>
<th>Grade 0</th>
<th>Grade 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Grade 1</td>
<td>Grade 4</td>
</tr>
<tr>
<td>Grade 2</td>
<td>Grade 5</td>
</tr>
</tbody>
</table>

Papilledema
(Increased Intra-cranial pressure)

- Headache
- Nausea
- Vomiting
- Diplopia (Abduction deficit – CN VI)
- Pulsatile tinnitus
- Transient Visual Obscurations (TVOs)
  - Last few seconds (uni or bi-lateral)
  - Transient ONH ischemia
- Optic Disc Edema
  - Bilateral/Asymmetric (anatomic difference in lamina)
  - RARELY Unilateral

Papilledema Work-Up

- MRI (r/o mass)
- MRV (r/o venous sinus thrombosis)
- LP - only after MRI (r/o meningitis / infection)
- Could be caused by mass of spine
- Idiopathic Intracranial Hypertension is a Diagnosis of Excusion

DISC DILEMMA

- IF THIS IS UNILATERAL DISC EDEMA, WHAT IS THE CAUSE?
**AGE**

<table>
<thead>
<tr>
<th></th>
<th>NAION</th>
<th>AION</th>
<th>OPTIC NEURITIS</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>AGE</strong></td>
<td>55-65</td>
<td>65-75</td>
<td>(60+)</td>
</tr>
<tr>
<td><strong>FEMALE/MALE</strong></td>
<td>3:1</td>
<td>3:1</td>
<td></td>
</tr>
<tr>
<td><strong>RACE</strong></td>
<td>LESS IN AA</td>
<td>LESS IN AA</td>
<td></td>
</tr>
</tbody>
</table>

**HEMORRHAGE**

<table>
<thead>
<tr>
<th>HEMORRHAGE</th>
<th>NAION</th>
<th>AION</th>
<th>OPTIC NEURITIS</th>
</tr>
</thead>
<tbody>
<tr>
<td>YES (72%)</td>
<td>YES/ NO</td>
<td>RARE</td>
<td></td>
</tr>
</tbody>
</table>

| DISC EDEMA | Diffuse 75% Focal 25% Pink Lux perf Art Vasoc ? | Diff edema Pallid (50%) | Diff edema Pink |

**VISUAL LOSS**

<table>
<thead>
<tr>
<th>VA</th>
<th>NAION</th>
<th>AION</th>
<th>ON</th>
</tr>
</thead>
<tbody>
<tr>
<td>49% = 20/64</td>
<td>34% = 20/200</td>
<td>21% = NLP</td>
<td></td>
</tr>
<tr>
<td>NADIR</td>
<td>Days (3.4)</td>
<td>Days (4.7)</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>PAIN</th>
<th>NAION</th>
<th>AION</th>
<th>OPTIC NEURITIS</th>
</tr>
</thead>
<tbody>
<tr>
<td>RARE</td>
<td>YES</td>
<td>78.8%</td>
<td>Precedes visual loss 92.2%</td>
</tr>
</tbody>
</table>

| Transient Monocular Blindness | NO | YES | NO |
NAION PREDISPOSING FACTORS

- SMALL, CROWED OPTIC DISC (DISC AT RISK)
- HYPERTENSION
- DIABETES
- CHOLESTEROL
- GENETIC
- SLEEP APNEA

GCA SYMPTOMS

- > 50 YEARS OLD
- HEADACHE
- TENDER TEMPORAL ARTERY
- MYALGIA/ ARTHRALGIA
- FEVER
- WEIGHT LOSS (Deceased Appetite)
- JAW CLAUDICATION
- MALAISE

Types of Optic Neuritis

- Perioptic Neuritis
- Neuro-Retinitis
- Papillitis
- Retrobulbar Optic Neuritis

PERIOPTIC NEURITIS

- SWOLLEN DISC
- ASYMPOMATIC PATIENT
- GOOD VISUAL FUNCTION
- EXUDATIVE
- SARCOID, SYPHILIS

NEURORETINITIS

- ACUTE VISUAL LOSS
- DISC SWELLING
- PERIPAPILLARY EXUDATIVE RD
- MACULAR STAR IN 2 WEEKS
- EXCELLENT PROGNOSIS
- NOT RELATED TO MS!
NEURORETINITIS

- SARCOID
  - (ACE / CXR, etc.)
- LYME DISEASE
  - (Lyme Titer / Lyme Western Blot IgG and IgM)
- TOXOPLASMOsis
- SYPHILIS
  - (RPR / FTA-ABS)
- CAT SCRATCH DISEASE
  - (Bartonella Quintana / Bartonella Henselae)

Optic Neuritis

- Papillitis
- Retrobulbar Optic Neuritis

Optic Neuritis – Papillitis & RON

- Young adult
  - More likely a woman
- Unilateral visual loss
- Progresses over hours to days
- 90% with pain, with eye movement
- Can be idiopathic, associated with MS, or other systemic conditions

SELF ASSESSMENT / REVIEW

Which optic disc appearance is most consistent with a presentation of acute optic neuritis?

a.  

b.  

c.  

d.  

(a, b, c, d)
This patient LEAST likely has?

a. Idiopathic intracranial hypertension
b. Optic disc drusen
c. Transverse venous sinus thrombosis
d. Meningitis

This patient has a prominent spontaneous venous pulsation in the left eye, and no visual complaints. What is the most likely diagnosis?

a. Papilledema
b. Buried optic disc drusen
c. Optic neuritis
d. Ischemic optic neuropathy

This patient most likely has?

a. Idiopathic intracranial hypertension
b. Sub-dural hematoma
c. Optic neuritis
d. Arteritic anterior ischemic optic neuropathy

This patient most likely has?

a. Giant cell arteritis
b. Sarcoid
c. Syphilis
d. Papilledema

This patient LEAST likely has?

a. Vitamin B 12 deficiency / folate deficiency
b. Pituitary adenoma
c. Multiple sclerosis
d. Bilateral occipital lobe infarcts

CASE 1
46 year old woman

- Noticed decreased vision OU x 3 months
  - Getting progressively worse
- Color vision is altered –blue and orange haze
- Denies eye pain or headache
- Sys Hx: Asthma
- Past ocular hx: unremarkable (past records retrieved – VA was normal one year prior)

Social Hx:
- Smoking 2 packs of cigarettes per day x 30 years
- Drinks heavily ("alcoholic" x 2 years)
- Admits to poor nutritional habits associated with her alcohol use

BCVA: OD 20/60 and OS 20/50
- Color 14/14 OD and 14/14 OS
- PERRL (-) APD
- CF: full OU
- HVF: general reduction of sensitivity and reduced foveal threshold bilaterally
- SLE and IOP normal OU

Labs for Optic Neuropathy

- CBC with differential and platelet count
- C-reactive protein (inflammation)
- ESR (inflammation)
- Lyme titer (if + get Western blot IgG and IgM)
- ANA with reflex titer (auto-immune disease)
- ACE (sarcoid)
- RPR (syphilis)
- FTA-ABS (syphilis)
- Vitamin B 12 (nutritional)
- Folic acid (nutritional)
- Methylmalonic acid (nutritional)
- Homocysteine (nutritional / inflammatory)
- SPEP (Serum Protein Electrophoresis) (possible malignancy)
Labs for Optic Neuropathy

- CBC with differential and platelet count
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- Methylmalonic acid (nutritional)
- Homocysteine (nutritional / inflammatory)
- SPEP (Serum Protein Electrophoresis) (possible malignancy)

Lab Results

- Low folic acid at 2.94 (normal > 5.4)
- Low normal vitamin B 12 at 464 (normal 200-1100)
- Normal methylmalonic acid
- Elevated homocysteine at 53 (normal up to 13.9)

Homocysteine

- Amino acid in the blood
- Increased with vitamin B12 or folate deficiency, genetic causes or renal disease
- Related to greater risk of cardiovascular disease, stroke, peripheral vascular disease
- Elevated levels = atherosclerosis
- Folic acid and vitamins B6 and B 12 break down homocysteine in the blood
- A 3umol/L drop in homocysteine = 16% less chance of heart attack, 24% less chance of stroke, and 25% less chance of DVT
- Levels above 14 umol increase risk of Alzheimer’s by 150%

Methylmalonic acid

- Early indicator of (occult) vitamin B 12 deficiency, or renal insufficiency
- Level is elevated when abnormal
- Methylmalonyl co A \( \rightarrow \) succinyl coA
  \[ \text{vit B12} \]

If both MMA and Homocysteine elevated = Vit B 12 Def
If MMA normal and Homocysteine elevated = Folate Def
Either can cause megaloblastic anemia

Toxic / Nutritional Optic Neuropathy

- Started on 1 mg Folic acid po qd
- Began taking multivitamins
- Need to d/c alcohol use, and improve eating habits
- Hematology consult requested to see if additional treatment is needed
  - IM/IV folic acid
  - High dose Thiamine

Pt refused alcohol support groups / counseling
Gave pt phone number for alcohol rehabilitation program
CASE 2

62 year-old woman

• Referred because of optochoroidal shunt vessels
• History of glaucoma
  – Using Travatan and Azopt OU
• No eyecare x 1.5 months due to lack of insurance
  – PCP did refill drops during that time

• Chief complaint
  – Blur with prolonged reading
  – Rare headaches from lack of sleep

• Systemic History:
  – Diabetes
  – Hypertension
  – Hypercholesterolemia
  – Rheumatoid Arthritis

• BCVA: OD 20/20  OS 20/20
• Color: OD 12/14  OS 14/14
• PERRLA (-) RAPD
• Normal ocular motility
• SLE: only mild lens changes
• TA: OD 17 mm Hg  OS 18 mm Hg
• BP: 142/80
• Pt is of Haitian descent
  – Came to US 6 years ago

• She recalls that in Haiti about 20 years ago
  – Decrease in vision in OS
  – Unsure of diagnosis, but remembers being told about some bleeding in the eye

CASE 3

73 year-old man

• CC: Has noticed a gradual decrease in vision OS
• Occasional headaches, longstanding, stable, attributes to sinus issues
• Systemic History
  • Diabetes
  • Hypertension
  • Hypercholesterolemia
  • Hyperthyroidism – uncontrolled
• Ocular History
  • Normal Tension Glaucoma x 7 years
  • Cataract Surgery OU
Exam Findings
- VA: 20/25 OD, 20/40 OS
- Color: 6/14 OD, 7/14 OS
- PERRLA (+) RAPD OS
- EOMs: no restrictions
- No ptosis or proptosis

Exam Findings
- Normal slit lamp exam
- IOP: 17 OD, 16 OS
- BP: 169/95
- DFE: 0.6 x 0.6 cupping OD
  0.75 x 0.75 cupping OS
- (-) edema

Assessment: Bitemporal hemianopia, suspect structural abnormality in area of suprasellar cistern

Plan: Order MRI of brain with and without contrast
Order pituitary gland function tests (prolactin, growth hormone, cortisol, TSH, FSH, LH, and IGF-1)
Normal Tension Glaucoma

- Diagnosis of Exclusion
- Many things can mimic NTG
- Especially if there is optic disc pallor and the VF does not match the ONH, additional work-up is needed
  - Lab testing to rule out infectious, inflammatory, nutritional
  - Neuro-imaging to rule out mass, abnormal enhancement

CASE 4

18 year-old girl

- CC: diplopia at D and in R gaze
  - Onset 2½ months prior
- Headache, constant
  - Onset 2½ months prior
  - No improvement with OTC medications
- Pain in right eye

Labs for Optic Neuropathy

- CBC with differential and platelet count
- C-reactive protein (inflammation)
- ESR (inflammation)
- Lyme titer (if + get Western blot IgG and IgM)
- ANA with reflex titer (auto-immune disease)
- ACE (sarcoid)
- RPR (syphilis)
- FTA-ABS (syphilis)
- Vitamin B 12 (nutritional)
- Folic acid (nutritional)
- Methylmalonic acid (nutritional)
- Homocysteine (nutritional / inflammatory)
- SPEP (Serum Protein Electrophoresis) (possible malignancy)
- Gave birth about 3 months prior
- Began noticing symptoms around that time
- Patient thinks symptoms began after getting epidural anesthesia
- Patient has “not felt right” since
- Very tired, “wants to sleep a lot”

- S/ P 3 Pregnancies
  - Full term birth January 2006
  - Elevated blood pressure
  - Terminated Pregnancy
  - Premature birth (at 34 weeks) October 2007
- Systemic Hx: Otherwise unremarkable
- Medications: Depo Provera (medroxyprogesterone IM)
- Report history of “lazy eye” (R lid droop)
- Denies any other health problems
- Denies trauma

**Examination Results**

- VA: OD 20/20  OS 20/20
- Color (Ishihara): OD 14/14  OS 14/14
  - No red desaturation
- Pupils: PERRL (-) APD
- CF: Full OU
- Exophthalmometry: OD 17 mm OS 18 mm
- Normal anterior segment exam
- IOP: OD 12 mm Hg   OS 12 mm Hg

**Ocular Motility**

- OD
  - 100
  - 100
  - 70
  - 100

- OS
  - 100
  - 100
  - 100

- NEGATIVE FORCED DUCTION TEST
CASE 5

49 year old woman

• Awoke 2 days prior, and noticed vision in right eye was not clear
  – Sudden change
  – Stable since onset
  – (-) eye pain
  – (+) headache, occasionally, stress-related
• SYSTEMIC HEALTH:
  – Diabetes x 20 years (last HbA1c: 11%)
  – Hypertension x 30 years
  – Hypercholesterolemia
  – Asthma
  – Sleep Apnea (CPAP broken)
  – Depression
  – s/p right mastectomy for breast cancer 7 years prior
    • Radiation and chemotherapy

• MEDICATIONS:
  – HCTZ
  – Lotrel
  – Novalog
  – Metformin
  – Naproxen
  – Singular

• OCULAR HISTORY:
  – Unremarkable

• SOCIAL HISTORY:
  – Unremarkable

• BCVA: OD 20/1000 and OS 20/20
  • Color 10/14 OD (with EF) and 14/14 OS
  • 50% reduced red saturation
  • 50% reduced brightness sense
  • Pupils – isocoric, 1.5 log RAPD OD

• Confrontation Fields:
  – OD: inferior nasal defect extending into inferior temporal and superior nasal quadrants
  – OS: possible mild inferior nasal defect

• HVF: poor reliability, significant VF loss OD

• No ptosis or proptosis

• Normal ocular motility examination

Poor reliability
• Slit Lamp Exam: Mild lens changes
• TONOMETRY: OD-12 mm Hg, OS- 13 mm Hg
• BP: 170/100
• Pulse: 69 bpm

DDX
• NA-AION
  — Consistent Features:
  • Sudden onset
  • Disc edema with hemorrhages
  • Disc at risk in fellow eye
  — Risk factors:
  • Hypertension
  • Diabetes
  • Sleep apnea

Follow-Up 2 months later
• Scheduled pt to see endocrinologist
• Scheduled for sleep study (needs new CPAP machine)
• Since last visit, increased medication for HTN and DM
• Some improvement in vision
1 year later.....

- Patient missed previously scheduled follow-ups

- Returns now due to vision loss in left eye!

- Blur OS x 6 days
- Went to ER – BP was high
  - Increased dosage of Clonidine
  - Pt put back on aspirin
    - Discontinued prior to breast reconstruction after breast cancer
- Prior to loss of vision
  - Not taking aspirin
  - Not wearing CPAP (says it is too tight)

- Labs done a few days ago
  - HbA1c: 11.6%
  - Cholesterol: 215
  - Triglycerides: 208

  - Pt states difficult to take care of herself because she is under a lot of stress

- BCVA: OD 20/200 and OS 20/125
- Color 11/14 OD and 4/14 OS
- Pupils – isocoric, 0.6-0.9 log RAPD OS
- Humphrey VF: unreliable
- Normal efferent function
• Slit Lamp Exam: Mild lens changes
• TONOMETRY: OD: 10 mm Hg, OS: 10 mm Hg
• BP: 115/64

Pt referred to endocrinology
• Pt educated on importance of controlling DM, BP etc
• Pt educated on need to use CPAP machine
• Pt educated on need to reduce stress

F/U 2 months later...

Avg RNFL thickness
OD: 58 um
OS: 328 um

Avg RNFL
Thickness:
OD: 60 um
OS: 96 um

CASE 6
40 year old asymptomatic woman

- Hx of keratoconus
- Came in for CL eval, and anterior uveitis was noted
- On Pred Forte qid OS and Nevanac (NSAID prodrug) qid OS
- Photosensitivity – stable x years
- Denies other symptoms

Systemic Hx:
- Hypercholesterolemia – no meds
- S/p removal of axillary lymph nodes (infxn?)
- S/p 2 full term uncomplicated pregnancies
- Asthma
- Meds: Advair, Combivent, Zyrtec, Claritin

VA: OD 20/25  OS 20/60
- Color: 13/14 OD and 0/14 OS
- PERRL (+) >1.8 log RAPD OS
- No ptosis or proptosis
- Normal ocular motility exam
- SLE: only few residual cells
- TA: OD 18 mmHg OS 21 mm Hg
- Normal neurologic exam

Labs for Optic Neuropathy
- CBC
- C-reactive protein
- ESR
- Platelet count
- Lyme titer (if + get Western blot IgG and IgM)
- ANA with reflex titer
- ACE
- RPR & FTA-ABS
- Vitamin B 12
- Folic acid
- Methylmalonic acid
- SPEP
Work-Up

- MRI – brain & orbits w & w/o gad
  - Arnold Chiari I malformation
  - No other structural abnormalities or abnormal enhancement
- Labs
  - CBC **elevated eosinophils 11.6%**
  - Platelets
  - ESR **H 85 mm/hr**
  - CRP **H 1.22**
  - Folate
  - B12
  - Lyme titer
  - RPR / FTA-ABS
  - ACE **H 254**
  - ANA

Chest CT

- Findings compatible with longstanding sarcoidosis
  - Bulky lymphadenopathy
  - Parenchymal changes
  - Scarring
- Lower lobe infiltrate? Possible superimposed bacterial pneumonic component
- Pt referred to pulmonologist and surgeon
  - Lung biopsy performed (+) for sarcoid

Neurosarcoid

- Occurs in 5-15% of pts with sarcoid
- CN VII is commonly affected
- CN II and VIII also affected
- Can be a mononeuropathy, peripheral nerve involvement, CNS involvement
- Can see leptomeningeal enhancement
- Active inflammation responds very well to steroids **(Oral steroids are fine for sarcoid)**

CASE 7

20 year-old man

- Pain OD on upgaze x few days
- Today, vision OD is “off”
- Denies diplopia, transient vision loss
- Denies headache
- A few days ago, he felt feverish, but did not check his temperature
- Fam HX: Father dx with Lupus in 20s
• BCVA: OD 20/50 and OS 20/20
• Color 14/14 OD and 14/14 OS
• Pupils – pharm dilated
• CF: full OU
• HVF: essentially normal OU
• SLE and IOP normal OU
• BP: 104/70
• Temp: 98.8 degrees

Humphrey Visual Field: At initial presentation. Note only slightly enlarged blind spot in the right eye, and fairly preserved central visual field.

• Pt denies any rashes (only when asked)
• He does admit to a scratch by a cat (kitten) several weeks ago (only when asked)
• A few weeks ago, his right eyelid was swollen
• Pt has several scars on his forehead, above right eye, and on his nose

Labs Ordered – told to have done today!
• CBC
• C-reactive protein
• ESR
• Platelet count
• Lyme titer (if + get Western Blot Lyme IgG and IgM)
• ANA with reflex titer
• ACE
• RPR
• FTA-ABS
• Bartonella Quintana titer
• Bartonella Henselae titer

Follow-up 5 days later
• Pt notes a spot in right vision, that is getting larger
• Reduced central vision
• Since last visit, has had chills and fever
• Has also had headache
• Decreased appetite
• Unable to work – doesn’t feel right
• Labs not done until 2 days ago – not complete
Lab results (so far)

- ANA (+) titer and pattern not yet known
- Lyme titer is (+) WB IgG (-), IgM (+)
- ACE slightly elevated at 70
- ESR: 44
- CRP: pending
- Bartonella titers: pending

• BCVA: OD 20/200 and OS 20/20
• Color 1/14 OD and 14/14 OS
• Pupils – trace RAPD OD
• CF: very large blindspot OD
• HVF: large blindspot OD – extending past fixation and superiorly out to 10 degrees
• SLE and IOP normal OU (-) cells

Humphrey Visual Field: 6 days after initial presentation. Note significant increase in blind spot in the right eye. The left field was unreliable due to patient fatigue.

Follow-Up: 6 days after initial presentation

Need to r/o Lyme, sarcoid, auto-immune disease
- Ds DNA (-)
- Repeat ACE (-), CXR (-)
- LP (-) for Lyme, sarcoid
• Bartonella titers
• Bartonella Quintana (-)
• Bartonella Hensalea (+)
• (+) IgG > 1:2560
• (+) IgM > 1:800

• DX: Cat-scratch Disease

Follow-Up: 15 days after initial presentation

Neuro-retinitis
• Cat-scratch
• Sarcoid
• Syphilis
• Lyme
• Toxo
• NOT typical in MS

Cat Scratch Disease
Typically transmitted by a kitten (by a scratch or a lick)

Only a minority of the exposures to B. Henselae result in cat-scratch disease. The ability of the cat to transmit the disease is transient

Most cases occur in fall / early summer - related to kitten births and flea infestations

80% of cases occur in patients under age 21

Starts with local infection, then lymphadenopathy, and rarely progresses – eg. Neuroretinitis, etc.

• Treatment:
  – Antibiotics
    » Doxycycline (pt vomited every time he took this medication)
    » Rifampin
    » Bactrim prescribed in place of Doxycycline
    » Pt then switched to Azithromycin by Infectious Disease
Cat Scratch Disease

- **Treatment / Response:**
  - Excellent prognosis - Most cases are self-limiting and fully resolve, even when involving the CNS
  - Drugs of choice – Bactrim, Gentamicin, Ciprofloxacin, Rifampin, Azithromycin

CASE 8

28 year old woman

- 2 weeks prior, noticed blurry vision OS
- Blur persists
- No pain in the left eye
- 3 days prior, noticed pain on eye movements OD
- 1 day prior noticed decreased vision inferior OD
- She is now having difficulty functioning

Follow-Up: 34 days after initial presentation

Humphrey Visual Field: 34 days after initial presentation. Note reduction in blindspot size in the right eye corresponding with a reduction in optic disc edema. A central scotoma persists in the right eye, related to the macular star and the reduced visual acuity.
• OTHER SYMPTOMS:
  – Weakness and tingling left thigh
  • Several episodes over past few years
  • Thought to be sciatica
  • Thought to be related to being over-weight
    – Lost 30 pounds, but symptom still recurred

• SYSTEMIC HEALTH:
  – asthma

• MEDICATIONS:
  – Albuterol, Advair

• OCULAR HISTORY:
  – Unremarkable

• SOCIAL HISTORY:
  – unremarkable

• BCVA: OD 20/25 and OS 20/40
• Color 14/14 OD and 10/14 OS
• 25% reduced red saturation OS
• 25% reduced brightness sense OS
• Pupils – pharm dilated
• Normal ocular motility exam

• Slit Lamp Exam: normal OU
• TONOMETRY: OD-16 mm Hg, OS- 16 mm Hg
• BP: 132/92
DDX

- Optic Neuritis OD, Optic Neuropathy OS
- ? Demyelinating Disease

WORK-UP
- Lab testing
- MRI brain and orbits with contrast
- MRI spine
- Possible lumbar puncture

TREATMENT:
- IV steroids x 3 days

RESULTS

- MRI brain and orbits – mild enhancement of posterior right optic nerve
- MRI spine: lesion at C3 level, but no brain lesions
- Pt diagnosed with Multiple Sclerosis at hospital

FOLLOW-UP

- Eye pain returns (was better on steroids)
- Severe headaches
- Leg paresthesias recur (better on steroids)

- BCVA: OD 20/25 and OS 20/25
- Color 14/14 OD and 10/14 OS
- Pupils – equivocal RAPD OS
- Normal ocular motility exam
• Because vision did not improve significantly after steroid treatment, and there was spine involvement but no brain involvement, need to consider:
  
  • NMO (neuro-myelitis optica or Devic’s disease)
  • Order NMO antibody testing (NMO-IgG Ab)
  • Also test for other causes of optic neuritis
  • (ANA, Lyme, ACE, FTA-ABS, etc)

• NMO (AQP4) antibody was POSITIVE
• Positive ANA, positive Sjogren’s antibodies

• Pt started on Azathioprine
• Then put on Rituximab  
  → some improvement in field of right eye

5 WEEKS LATER

Neuro Myelitis Optica (NMO)

Neuromyelitis optica is, like Multiple Sclerosis an inflammatory, demyelinating syndrome of the central nervous system

Within 5 years of disease onset, more than 50% of patients with relapsing neuromyelitis optica are blind in one or both eyes or require ambulatory help.

The detection of neuromyelitis optica immunoglobulin G (NMO-IgG), an autoantibody, in the serum of patients with neuromyelitis optica, distinguishes neuromyelitis optica from other demyelinating disorders.

NMO-IgG binds to aquaporin 4, which is the main channel that regulates water homoeostasis in the central nervous system.


Using typical MS treatments for NMO can cause NMO exacerbation!
- Interferon-beta
- Natalizumab
- Fingolimod

Thank you.