Round 2: Neuro, Optic Nerve, & Orbit Brainteaser Cases

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Same rules apply as before...

- I have no disclaimers or conflict of interests to report.....
- Don't look ahead!
- I'm not perfect...
- Some cases are more straight forward than others...
- I will email you my reference list if you want it....

- Email me: cborgman@sco.edu

Case History...

- 41 YO AAF
- PMH: (+)HTN, (+) DM, (+)CVA 7 years earlier with L-sided weakness
- BCVA 20/25 OD, 20/25 OS
- (+)APD OS
- CVF = very constricted
- SLE = WNL OU
- IOP = 17 OD, 16 OS
- DFE = see pics

Case #1: “Doc, I can’t see much.”

DFE...

OCT

Avg RNFL:
OD 53 μm
OS 66 μm
Pt Outcome...

- Ordered MRI
- MRI confirmed right optic tract lesion
  - Longstanding lesion per report

- LV Referral

- Bottom Line:
  - If 24-2 is “grap” then consider trying 10-2 after

Case #2:

“Doc, I see double…”

Case History...

- 30 year old white female
- New onset diplopia
- Worse when looking to left side
- (-) HA, (-) trauma
- No systemic, ocular history
- No medications
- (+) pregnant

Exam...

- VA = 20/20 OD, 20/20 OS
- Pupils = PERLA, (-) APD
- EOM’s = see figure; (+) nystagmus OS in left gaze
- CVF = WNL  →  HVF WNL OU too
- Adnexa/SLE = WNL OU
- IOP = 12 mmHg OU
- DFE= WNL OU
Dx = Internuclear Ophthalmoplegia...

- Lesion = ipsilateral MLF on side where adduction is absent
- Abducting nystagmus in fellow eye

Background...

- Most common causes:
  - MS
  - Stroke/CVA
  - Tumor
  - AVM...

- Imaging Pearl: Focus MRI imaging at level of pons
- BINO = both MLF’s implicated; (-)adduction OU
- WEBINO = subtype of BINO, midbrain location given no convergence

Is Convergence Important??...Yes!

- (-)Convergence = midbrain lesion
- (+)Convergence = pons lesion

Final Diagnosis...

- Dx = Right INO secondary to Multiple Sclerosis
  - Convergence was intact too! Lesion = PONS!

- IV steroids given by neurology

- INO resolved on its own in 2 months...

- Regular follow up with Neurology to monitor MS
  - At last follow up → neuro deciding on which MS medication to start with

Case #3:

"Doc, sometimes I see double..."

Case history...

- 50 year old white male
- First eye exam in 8-10 years
- History of brainstem stroke 8-10 years ago
- Eyes bothered him at first but no issues anymore
- MRI with neurologist at the time (8-10 years ago) found something, but he can’t remember what...
- “People say my eyes look funny...”
- Only here because wife made him make appointment...
Exam...

- VA = 20/20 OD, 20/20 OS (cc)
- Pupils: WNL OU
- CVF = WNL OU
- EOM's: see pictures
- SLE: WNL OU
- IOP = 18 mmHg OU
- DFE = WNL OU

Diagnosis...

- Left Gaze Palsy OU + Left INO = One-And-Half Syndrome
- Pontine lesion (MS, CVA, mass) $\rightarrow$ new onset needs brainstem MRI with contrast, concentrated on Pons
- In old OAHS $\rightarrow$ monitor, control underlying systemic issues
  * Rewire if any worsening occurs
- Lesion involves: bilateral MLF's $+$ one of the CN VI (Abducens) nerves

Case #4:

“Doc, I see double…”

Case History...

- 62 year old white male
- Referred from ER after falling down stairs at home and being found unconscious 1-2 days ago
- CC = double vision, worse right gaze, since regaining consciousness
- CT and MRI = CVA/Stroke of brainstem
- Left sided/cranial palsies since fall
- Right Facial Droop/Palsy
- Hearing unaffected (CN VIII), facial sensation (CNV) unaffected
- PMH: (+)HTN, (+)Hypercholesterolemia, (+)DM
Exam...

- VA = 20/25 OD, 20/25OS cc
- Pupils: PERRLA, (-) APD, (-) anisocoria
- CVF: WNL OU → HVF-WNL OU
- EOMs: see figure
- Adnexa: Right-sided facial droop
- SLE: WNL OU
- IOP = 18 mmHg OU
- DFE: WNL OU

Right Gaze Palsy + Right Side Facial Droop

Clinical Summary...

1. Right gaze palsy
2. Right-sided facial droop
3. Left-sided hemiparesis

- Lesion = inferior pons!

CN VII Review...Forehead is key!

A. Lower motor neuron lesion = half of face involved
   - Nuclear/fascicle nerve lesion
B. Upper motor neuron lesion = lower 1/4 face involved
   - Supranuclear lesion

- Forehead is the key!

- My patient can be localized to lower motor neuron based on total right side of face involvement.

Right Gaze Palsy 2° CVA...

- Inferior Medial Pons = location of lesion

What about the left sided hemiparesis?

- Decussation of corticospinal tract = upper medulla!
Pt’s Diagnosis...

1. Ipsilateral CN VI Palsy
2. Ipsilateral CN VII Palsy
3. Contralateral hemiparesis

- Lesion = ventral paramedian pons secondary to occlusion of right paramedian branch of the basilar artery/AICA

Millard-Gübler vs. Foville’s Syndrome?

- Ipsilateral CN VI or Ipsilateral Gaze Palsy
- Ipsilateral CN VII/Facial Palsy
- Contralateral hemiparesis
- Lesion = ventral paramedian pons
- Only V, VII, corticospinal tracts are involved

- Ipsilateral Abducent nerve or gaze palsy
- Ipsilateral CN VII/Facial palsy
- Horner’s Syndrome
- Analgesia of face (CN V)
- Peripheral deafness (VIII)
- Loss of taste (V, mandibular branch)
- Lesion = dorsolateral pons
- Only V, VI, VII, VIII are involved

Back to patient...

- Follow-up 4 weeks later:
  - Drastic improvement in Sn/Sx over 4 week timespan
  - Still had some facial weakness and hemiparesis but diplopia had resolved entirely

Case #5:

- “Doc, my neurologist wanted me to see an eye doctor.”

Case History...

- 59 YO AAF
- LEE 3 years ago
- ALL: NKDA
- Meds: Lisinopril, HCTZ, Atenolol, Simvastatin
- PMHx:
  - Hypertension x 12 years reported as “controlled” with medication
  - CVA: May 2015 and January 2009
  - Right sided weakness since stroke

Initial Exam...

- BCVA = 20/25+ OD, 20/30- OS
- Pupils: WNL
- CVF: WNL
- EOM’s: see pics...
- SLE = grossly normal
- DFE = see pics...
- IOP = 19 OD & 18 OS
Dilated Fundus Exam

- Posterior Segment:
  - Optic Disc: Round, Distinct, +1 Round OD
  - Retina: Cotton Wool Spots OD
  - Macula: Flat & Intact
  - Posterior Pole: Cotton Wool Spots (+) Hemes OD
  - Vessels: Crossing Changes OD

Considerations???

- Is there more than one location???
- Is there any one location that could cause all of the symptoms?
  - CVA occurring at/near the level of the pontomedullary junction where both CNVI and CNVII origins arise, including MLF, abducent nucleus, medial lemniscus and facial colliculus
- MRI results requested in plan to confirm suspected lesion(s) location

MRI Results

- Axial T2-weighted image showing a slightly hyperintense signal in left lower tegmental pons (yellow arrow)

MRI Results

- Axial Diffusion-Weighted MRI scan showing obvious hyperintense lesion at level of left inferior tegmental pons (yellow arrow)

MRI Results

- Axial Attenuated Diffusion Coefficient (ADC) weighted image showing hypointense signal in left inferior tegmental pons (yellow arrow)
Lesion in Left Lower Pons (Transverse Cross Section)

What about the contralateral hemiplegia???

- Corticospinal fibers are responsible for voluntary limb movement
  - Origin: motor cortex of precentral gyrus in frontal lobe
- Decussation fibers = upper medulla!
  - Inferior to lesion in this case!
  - Left pons lesion would lead to right-sided hemiparesis as in this case!

Quick Re-cap...

- **4 components:**
  1. Contralateral Gaze Palsy & Internuclear Ophthalmoplegia
     - Aka: One-And-A-Half Syndrome (OAAH)
  2. Ipsilateral Peripheral Facial Nerve Palsy (CN VII)
     - Marked hypertelorism, reduced blink rate, and weakened lid closure
  3. Contralateral Hearing Impairment (CN VIII)
  4. Contralateral hemiparesis/hypoesthesia
     - Paralysis or limb weakness opposite the body of suspected brain infarction/lesion

$$1 \frac{1}{2} + 7 + 8 + \frac{1}{2} = 17 \rightarrow \text{“Seventeen” Syndrome}$$

Causes/Lesions of OAAH Syndromes

- Vascular
- Demyelinating
- Tuberculosis
- Cystercerosis
- Endocarditis
- Systemic Lupus Erythematosus
- Giant Cell Arteritis
- AV malformation
- Trauma
- Aneurysm
- Tumors

Stats of One-And-A-Half Syndromes

- **8%** of all pontine lesions cause OAAH syndromes
- Of all OAAH syndromes:
  - 76% have additional cranial nerve involvement
  - 15% have 5th Nerve/TG (gemininal) involvement
  - 30% have 7th Nerve/Facial involvement
  - 10% have 8th Nerve/Vestibulocochlear involvement
  - 5% have Horner Syndrome
  - 30% have contralateral hemiparesis
  - 35% have contralateral hemihypoesthesia

OAAH Syndrome Spectrum...
1. OAAH + Facial Nerve = 8 ½ Syndrome
   • 1 ½ + 7 = 8 ½
2. OAAH + Facial Nerve + Hemiparesis = 9 Syndrome
   • 1 ½ + 7 + ½ = 9
3. OAAH + Facial Nerve + Trigeminal Nerve = 13 ½ Syndrome
   • 1 ½ + 7 + 5 = 13 ½
4. OAAH + Bilateral Facial Nerve Palsies = 15 ½ Syndrome
   • 1 ½ + 7 + 7 = 15 ½
5. OAAH + Facial Nerve + Vestibuloocular N. + Hemiparesis = 17 Syndrome
   • 1 ½ + 7 + 8 + ½ = 17

Additional Labwork to Consider if CWS Linger ...
• CBC c diff.
• CMP
• HIV
• RPR/FTA-ABS
• ANA
• Hepatitis Panel
• PT/PTT
• Sickleux
• ESR
• Chest X-ray

3 month follow-up exam...
• Nine Syndrome findings slightly improved but still present
• Cotton wool spots resolved with improved systemic control
• Therefore, no lab work ordered at this time
• Follow up x 6 months.

Case #6:
“Meh, I’m good but I feel clumsier than usual.”

Case History...
• 82 YO WM
• PMH: (+)Pacemaker installed, (+)asthma, (+)gallbladder removal, (+)DVT leg 4-5 years prior
• Meds: Benicar, Symbicort, Claritin
• Allergies: PCN
• (+)Phaco OU 1 year prior
• (+)blepharoplasty 5-6 years prior

Exam...
• BCVA 20/20 OD, 20/20 OS
• Pupils/EOM’s: WNL, (+)APD
• CVF: inf restriction OU
• SLE: WNL
• DFE: see pics
• IOP: 18 mmHg OU w/ Goldmann
• BP 126/69
Bilateral Cuneus Lobe Lesions of Occipital Lobes!

What happened to patient?

- Attempted to order neuroimaging...
  - (MRI due to pacemaker)
- Pt = "Off the Grid"
- Phone call/letters = unanswered...

What we expect to find on MRI/CT...

Case #7:

"Doc, it's hard for me to focus and I lose my place often when looking at things."

Case History:

- 60+ish YO AAF
- CC: hard to read and focus"
- PMH: HTN, hypercholesterol, (+)CVA several years earlier
- Meds: Lisinopril, HCTZ, Simvastatin
- ALL: NKDA
Exam:
- BCVA: 20/25+ OD, 20/30- OS
- EOM’s: FROM OU
- Pupils: PERRL, (-) APD
- CVF: “confusing”
- SLE: WNL OU, early cataracts OU
- DFE: WNL OU
- IOP: low teens OU

Visual Fields

Diagnosis???
- Checkboard Pattern Visual Field Defects!

What would we find on MRI?
- Left Cuneus Lobe Lesion AND Right Lingual Gyrus Lesion!!!

Case History
- 49 YO WM
- CC: “neurology wanted my eyes checked out”
- (+) Hemorrhagic & ischemic CVA 5 mo earlier
- 2’ myotic aneurysm
- (+) Endocarditis 5 mo earlier
- FHO: unremarkable
- Meds: ASA 325 mg, Levetiracetam, Citalopram, Metoprolol, Zolpidem, Pantoprazole, no amphotericin B
- Allergies: NKDA

Case #8:
“Doc, I’m lucky to be alive...”
Exam...
- BCVA = 20/20 OD, 20/20 OS
- Pupils = PERRLA (+)APD
- EOM's = Full OU
- Pursuits/Saccades = deficient/impaired
- CVF = Restricted inf temp OU
- SLE = Normal OU
- DFE = Normal OU
- IOP = 14 OD, 12 OS

Acute/Initial CT = Stroke

Brain Blood Supply Review...

Aneurysmal Coiling

Aneurysmal Coil on CTA (5 mo later)
5 mo later

T1 Axial
T1 Axial + C
T1 Axial
T2 Axial

6 mo later

When in doubt, go back to the case history!

Endocarditis

- Def: Infection of heart valves
- Age onset: 44-64 YO
- M:F (2:1)
- Highest risk in:
  - Drug addictions, drug users
  - Immunosuppressive therapy
  - Invasive procedures
  - Prosthetic heart valves
  - Diabetes
  - Mitral valve prolapse
  - Long-term catheterization
  - Malignancy

Endocarditis

- Aortic valve > Mitral valve > Tricuspid valve
- Infective Types:
  1. Bacterial (90-95%)
     - Staph, Aerues (70%), Staph, Pneumoniae, Strep, Pyogenes
     - 30% mortality rate
  2. Fungal (5-10%)
     - Candida (1-2%), Aspergillus (25-40%), Mixed (41%)
     - 80-94% mortality rate
- 50% cases suffer an embolic event
- 30% with cerebral embolization (CVA)
- 20% with embolic event at initial presentation
- Almost always requires heart replacement surgery

Fungal Endocarditis

https://en.wikipedia.org/wiki/Tricuspid_valve
https://en.wikipedia.org/wiki/Infective_endocarditis
Classic Signs of Endocarditis...

- Finger Clubbing (8%)
- Splinter Hemorrhages (nail beds) (7%)
- Osler nodes (finger tips) (4%)
- Janeway lesions (palms/soles) (4%)

- Roth's spots
  - 1-3%
  - Anemia, leukopenia, HTN, DM

- All from immune complex formation in those tissues

Where does the fungemia come from???

- Pneumonia
- Sinusitis
- Dental extraction
- Intracranial surgery
- Antibiotics and/or Immunosuppressants → Δ natural flora
- Abdominal/Gynecological surgery
- IV drug(s)

Endocarditis Diagnosis...

1. Blood Cultures
   - Usually consistent and low grade
2. Vegetation Culture
3. Valve histology
4. Echocardiograms
   - Transesophageal > Transthoracic
   - Visualization in 77-83% cases
   - Left > Right

Fungal Endocarditis MOA & Tx...

- **MOA**: Fungal endocarditis cannot occur unless the valve endothelium is prior traumatized (or congenital defect)
  - Fibroplatelet bed formation → fungal adhesion

- 80-94% mortality rate!
  - Prosthetic valve fungal endocarditis → 100% mortality rate!
  - Debridement, replacement, antifungal = 50% rate reduction
  - 1 year survival rate = 45-55% (surgery and antifungal)
  - 1 year survival rate = 36% (antifungal alone)
- DOC = Amphotericin B
- Fluconazole (second DOC)

Endocarditis “Downstream” Complications

- **Stroke** (6-31%)
- **TIAs** (27%)
- **Myotic aneurysms** (15%)
  - Aortic (3%)  
  - Visceral arteries (24%)  
  - Extremity arteries (22%)  
  - Intracranial (15%)
- Subarachnoid hemorrhage (3-7%)
- **Intracranial hemorrhage** (3-7%)
- **Seizures** (0.5-11%)
- Meningitis (0.3-16%)
- Headache (2.5-5%)

Mycotic Aneurysm

- **Def**: Aneurysm 2' infection of vessel wall
- **Oslerian MA**
  - Embolization from cardiac vegetations
- Hematogenous seeding
  - Bacteremia → seeding of arteries → weakened vessel walls → aneurysm formation
- Infected aneurysms
  - Aneurysm forms first → bacteremia invades aneurysm
- Others
  - Mechanical injury
  - Contiguous spread
Top 3 Modern Day Causes of MA

1. Trauma
2. Endocarditis (10-20% cases)
3. Bacteremia


Mycotic Embolization

- Cerebral and Femoral arteries → most common sites

- Cerebral:
  - MCA = most common site of cerebral embolization (57.4%)
  - 75% other MCA
  - PCA = 17.6%
- Fungal endocarditis → Mycotic Embolization in 30-55% cases
- Worst complication = mycotic aneurysm rupture → SAH → death

Bacterial Mycotic Aneurysms...

- Staphylococcus aureus  
- Streptococcus pyogenes  
- Salmonella  
- E. coli  
- Immuno compromised:
  - Campylobacter  
  - Listeria  
  - Mycobacterium tuberculosis

- Most common

Fungal Mycotic Aneurysms...

- Aspergillus species  
- Candida species  
- Most common fungi

- Overall
  - 3-15% of infected aneurysms
  - Mortality Rate = 84-100% !!!!
  - Immunosuppression!
  - Saccular or Fusiform shapes

Pt Summary...

- Pneumonia 1 month prior?  
  → Fungal vs. Long Duration of Abx Tx???
- Fungal Endocarditis → Candida albicans  
  → immunosuppression work up = negative
- Mycotic Aneurysm → Hemorrhagic CVA
- Inf Quad Defect Right Side & Left Sided Hemiparesis

Case #9:

- “Doc, I’m good to go… I just need new glasses.”
Case History

- 69 YO AAF
- CC: "Blurry vision"
- OD>OS, onset unknown, (+)trauma, (+)pain
- PMH: (+)HTN, (+)Type 2 DM
- Meds: Metformin, Lisinopril

- BCVA: 20/50 OD, 20/60 OS
- PCIOL OD, Nuclear Cataract OS
- IOP: 18 mmHg OU
**Neuroimaging...**

- Normal x 2 observers!
- I'm not so sure...atrophy???

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**How do we measure chiasmal atrophy?**

- N=123
- In 82% of cases, there was one image in which the optic chiasm and proximal optic nerves were clearly seen.
- **Coronal high-field T1-weighted** imaging = best
- "Our study suggests that optic chiasms with...a width between 10.6 mm and 17.4 mm (2 SD from the mean) can be considered definitely normal."

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**Horizontal Width of 10.6 – 17.4 mm**

- 16.4 mm
- 7.5 mm

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**What happened to patient?**

- Don't know...
- No-showed for follow ups...
- Has not returned phone calls/reschedule attempts...

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**Case #10:**

"Doc, I have a droopy lid, sort of dilated pupil, I see double when I lift my lid, and I'm having a hard time moving the right side of my body......also I'm having some trouble with my side vision."

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**History...**

- 72YO AAF
- Sudden onset diplopia
- Sudden onset visual field defect
- (trauma
- (+)DM, (+)HTN, (+) cholesterol
- FMI: (+)glaucoma—mother
- SH: +/smoker
- ALL: NKMA/NKDA
Exam:

- VA = 20/25 OD, 20/40 OS
- CVF = inferior left quadrant defects OU
- Pupils = 3/5 bright, 4/6 dark (+)APD
- EOM's = see pics
- Adnexa = see pics
- SLE: grossly unremarkable, except for cataracts OS>OD
- IOP = 13 mmHg OD, 14 mmHg OS
- DFE: unremarkable, (pallor, (ONH edema OU

Clinical Summary So Far...

1. Third Nerve/Oculomotor Nerve Palsy OS
2. Anisocoria (larger OS)
3. Right sided hemiplegia
4. Left Inferior Quadrantanopia OU

Wait a minute...What about the VF defect?

Dx = Weber's Syndrome...(Midbrain Lesion)

- Pupil involvement/Anisocoria?
- Left CN III Palsy
- Contralateral hemiparesis/hemiplegia
- How?
- Visual Field Defect?
  - Inf quad Defect localizes to Right Parietal Lobe or Right Occipital Lobe

5/19/2016
Hypothetical MOA Option #1 = **Two** Diagnoses...

1. Weber Syndrome
   - Ipsilateral CN III Palsy OS (could include anisocoria)
   - Contralateral/Right side hemiplegia/paresis

2. Left Inferior Quadrantanopic Defect OU
   - Second infarction of Right Parietal Lobe

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Hypothetical Option #2 = **One** Dx

- Hemorrhage in right parietal lobe can form hematoma
- Hematoma acts as a tumor shifting brain down and left
- Uncal herniation of temporal lobe through tentorial notch compressing CN III

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Patient needed convincing...

- Denied first 2 requests for previous MRI/CT records
- Agreed on 3rd attempt...

- Final answer = Option #1! (2 lesions total!)
  1) Right posterior parietal/anterior occipital lobe ischemic infarction
  2) Left thalamic infarct extending into left midbrain
  3) Multiple bilateral posterior cerebral artery stenoses; R>L

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Midbrain Blood Supply...

- Posterior Cerebral Artery & Its Branches

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Occipital Lobe Blood Supply...

- Posterior Cerebral Artery & Its Branches

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Thank you!!!