Neuro-ophthalmic Diagnoses
Not to Miss

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Importance of Neuro-ophthalmic Diagnoses

- Severe consequences
  - Irreversible damage to the patient (potentially treatable; things that can kill the patient)
  - Medicolegal implications

- Unusual unexpected pattern
  - Delay in diagnosis: expense
  - Consequence for patient
  - Do you have to call me every day?
Severe Consequences

- Keep you out of trouble
- "First we’ll kill all the lawyers"
- This is what you came to this lecture for
BAW 81yo female

9/10: ↓Va

Va 20/200, 2/200

N 20/400 OD

VF:

Ext: w/q

P: 1.2log RAPD

EOM: full

SLE: PC-IOL

Ta: 10 OU

Fundus:
BAW – POH

Glaucoma X 20yrs

2000: Phaco OD

2007: Phaco OS

9/10 (2wks before): check up glaucoma

Va 20/30 OU

Ta: 16 OU

9/10 (10d before): light sensitivity

Rx: “Wear sunglasses”
9/10 (9d before): “Pain in head & jaw, sometimes shoulder + sore throat”
BAW – W/U

F/A (9/10):
BAW – W/U
9/10: ESR: 74
Platelets: 554k
CRP: 77.9
HP 78yo male
PMH: AODM
POH: POAG on Timoptic
6/88: 3mo h/o temporal & occipital HA, jaw claudication
4d h/o blue vision
Evaluation: Va 20/50, 20/60
Ext: tender temporal artery
ESR: 75
HP - F/U

1d h/o dec Va OD

Va HM, 20/60; N 6pt OS

VF:
Ext: prom temporal artery

P: 1.2log RAPD

EOM: full

SLE: 2+NS

Ta: 13/17

Fundus:
HP - W/U

F/A:
HP - W/U

Ta Bx: GCA

Rx: 2gm IV methylprednisolone

Va 20/200,20/60

F/A:

80mg prednisone
CWA 86yo male
10/05: 1yr h/o dec Va OS
2d transient dec Va OD
Va 20/50, CF 1’
N J7, 20/800
VF:
P: LAPD
SLE: 3+NS
Fundus: ARMD
CWA – Rx

11/05: Phaco OS
CWA - F/U

12/05: episodic visual loss OD

Va CF 5’, HM; N J10 OD

VF: severe constriction

Ext: w/q

P: LAPD

SLE: 1+ C/F OS

Ta: 16/10

Fundus: disc edema OD

ESR: 24
CWA - F/U

1/06: ER w/ “vesicular” eruption

Dx: zoster

Rx: Valtrex

Residual tenderness over scalp

Trouble w/ swallowing
CWA

3/06: Referral

Va 20/50, LP

N 8pt OD

VF:

Ext:

P: >1.8 log LAPD

EOM: full

SLE: 3+NS OD

Ta: 13 OU

Fundus:
CWA - W/U

OCT:

<table>
<thead>
<tr>
<th>Parameter</th>
<th>OD</th>
<th>OS</th>
<th>Diff (OD-OS)</th>
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<tbody>
<tr>
<td>Total thickness (microns)</td>
<td>216</td>
<td>200</td>
<td>16</td>
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<tr>
<td>Macula thickness (microns)</td>
<td>137</td>
<td>127</td>
<td>10</td>
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<tr>
<td>Average Retinal Thickness (microns)</td>
<td>186</td>
<td>161</td>
<td>25</td>
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</table>

Patient/Scan Information

AUSTEN CHARLES
DOB: 3/21/1991, ID: 1895500, Male
Scan Type: Fast Macular Thickness Map
Scan Date: 3/7/2016
Scan Length: 5.0 mm
CWA - W/U

ESR: 17

CRP: 1.1
CWA - W/U

TA Bx: GCA
CWA - Rx

IV pulse steroids

Prednisone
Severe Consequences

- Giant cell arteritis
Giant Cell Arteritis

- Only 5% of AION
- ESR not always elevated
- Preceding amaurosis
- Diplopia possible
- Jaw claudication very suggestive
- Rx first (IV steroids); bx later
- Incidence dramatically inc w/ age
EL 82yo woman

7/85: 6wk h/o L brow ache

5wk h/o L ptosis + diplopia

4wk h/o sudden visual loss OS

Dx: “GCA”

Bx TA: negative

Rx: prednisone
EL- PE

Va 20/30, NLP

VF: slight constriction OD

Ext: 8mm L ptosis, H 19/22

P: 4+ LAPD

EOM: absent abd OS, limit vertical

SLE: 2+ NS

Fundus: early OA OS
EL - W/U

Review CT:
EL - W/U

FNAB:
JM 35yo male

10/96: 20d h/o redness L face
4d h/o dec sensation L face
2d h/o double vision

N 3pt, 5pt

VF: CF all quad

Ext: no corneal OS, H 15/16

P: 5/3 w/o APD

EOM: dec abd OS

Fundus: nl DMV
JM - PMH

Dx: DM

Ketoacidosis before transfer

Rx:

Timentin

Vancomycin

Acyclovir

Cipro

Amphotericin
JM - W/U

MRI:
JM - Rx

Maxillectomy + ethmoidectomy

Path: hyphae

Continue Amphotericin + Fluconizole
Severe Consequences

- Giant cell arteritis
- Infectious orbital apex syndrome
Orbital Apex Syndrome

• Mucormycosis/Aspergillosis
• Not always immune suppressed (acidosis)
• Acute visual loss: vascular involvement
• Paranasal sinus involvement (endoscopic)
DMD 54yo male

10/95: Awakens from CABG w/ no vision

CT: “negative”  Dx: “Occipital stroke”

Va: NLP OU; P: non reactive
Severe Consequences

- Giant cell arteritis
- Infectious orbital apex syndrome
- Pituitary apoplexy
Pituitary Apoplexy

- Acute onset
  - Decrease vision
  - Ophthalmoplegia
  - Mental status changes
- Pituitary tumor not always known before hand
- Surgery may precipitate a bleed
PE 45yo female

9/08: 1wk HA + diplopia

N 6pt,3pt

VF: full

Ext: R ptosis

P: 4.5/3 w/o APD

EOM: 

PLE: wnl

Tt: soft OU

Fundus: nl DMV
PE – W/U

MRA (9/08):
9/08: Coil P-com aneurysm
PE – F/U

12/08: Double gone

Va 20/20 OU

VF: full

Ext: Palp 7/9

P: 3/3.5 w/o APD

EOM:

SLE: wnl

Tt: soft OU

Fundus: nl DMV
3/09: Double vision

Va 20/20 OU

N 4pt,3pt

VF:

Ext: palp 8/10

P: 3-2 w/o APD

EOM:

SLE: wnl

Ta: 20/16

Fundus: nl DMV
GED – W/U

Pneumotonometry
GED – PMH

CTA:
Severe Consequences

• Giant cell arteritis
• Infectious orbital apex syndrome
• Pituitary apoplexy
• Aneurysmal IIIrd nerve palsy
Aneurysmal IIIrd

- Pupil sparing not present (for acute p-com)
  - Pupil sparing not rare w/ cavernous mass
- Potential for missing w/ MRA/CTA
- Sentinel bleeds
SFR 59yo female

9/99: 2-3 mo h/o ↓ Va OD X 1-2 min

Va 20/20 OU

N 3pt OU

VF:

Ext: w/q

P: w/o APD

EOM: full

SLE: trace NS

Ta: 18/17

Fundus:
SFR - W/U

MRI:
SFR - W/U

Angio:
SFR - Rx

10/99: R carotid endarterectomy
Severe Consequences

- Giant cell arteritis
- Infectious orbital apex syndrome
- Pituitary apoplexy
- Aneurysmal IIIrd nerve palsy
- Amaurosis fugax
Amaurosis Fugax

- Carotid artery disease (17% proximal vessels)
- Retinal emboli
- Risk of hemispheric stroke
  - Age >75
  - Male
  - Hx of hemispheric TIA
  - >80% carotid stenosis
  - Lack collateral circulation
- Non invasive carotid study
- Death from cardiovascular disease
NFL 43yo male

4/08: Transient ↓ Va OD

Va 20/20 OU

N 3pt OU

VF:

Ext: w/q

P: w/o APD

EOM: full

SLE: wnl

Ta: 15 OU

Fundus:
NFL – PMH

Very active: hang gliding, running

4/08: Dull R HA at time of visual loss (while hiking) persistent
NFL – W/U
NFL – W/

Cocaine test:
NFL – W/U
ODM: 90/45; 105/50
CT (4/08):
NFL – Rx
Heparin
Coumadin
6mo later switch to ASA
Severe Consequences

- Giant cell arteritis
- Infectious orbital apex syndrome
- Pituitary apoplexy
- Aneurysmal IIIrd nerve palsy
- Amaurosis fugax
- Carotid dissection
Carotid Dissection

- Traumatic (chiropractic) vs spontaneous
- Risk: fibromuscular dysplasia, Ehlers-Danlos IV

Symptoms:
- Facial pain
- Horner’s syndrome (58%)
- Dysgeusia

Consequence
- Hemispheric stroke
- CRAO/BRAO
Commonly Missed Diagnoses

- Afferent system: decreased vision
- Efferent system: double vision
- Orbital findings
CMB 65yo female

2/10: ↓Va

Va 2/200, 20/30

N 20/800, 3pt

VF:

Ext: w/q

P: >1.8log RAPD

EOM: full

SLE: PC-IOL OD, 2+NS OS

Ta: 14 OU

Fundus:
CMB – PMH

Breast cancer

s/p mastectomy

Diverticulitis

2007: Lymphoma
CMB – POH

2005: Cataract

10/05: Phaco OD

11/05: Va 20/25 OU

2/08: YAG OD

10/08: Va 20/25- OU

1/10: 1yr h/o ↓Va OD

Va CF, 20/25
CMB – W/U

OCT NFL (2/10):

RNFL THICKNESS AVERAGE ANALYSIS

<table>
<thead>
<tr>
<th></th>
<th>OD (N=3)</th>
<th>OS (N=3)</th>
<th>OD-OS</th>
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<tr>
<td>Tavg</td>
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<td>38.00</td>
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<tr>
<td>Favg</td>
<td>65.00</td>
<td>66.00</td>
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<tr>
<td>Avg.Thickness</td>
<td>52.47</td>
<td>46.79</td>
<td>5.68</td>
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Patient/Scan Information

burrell
catherine

DOB: 1/25/1945, ID: 2180316, Female
Scan Type: Fast RNFL Thickness (3.4)
Scan Date: 2/25/2010
Scan Length: 10.87 mm
CMB – W/U

B-scan (2/10):
CMB – Rx

3/10: Stent + coil RICA aneurysm
Commonly Missed Diagnoses

- Compressive optic neuropathy
Compressive Optic Neuropathy

- Usually slowly progressive
- Get the old records:
  - Amblyopia
  - Previous tumor
- “Chronic optic neuritis”
- Importance of visual fields
- Importance of an afferent pupillary defect
- Avoid attributing to other diseases
SEY 25yo male

8/07: ↓Va

Va 20/20, 20/40

N 3pt OU (5cm OS)

VF:

Ext: w/q

P: w/o APD

EOM: full

SLE: wnl

Fundus: nl DMV
SEY – POH

Glasses X 2yrs

2005: LASIK
SEY – W/U

Corneal topography:
Commonly Missed Diagnoses

- Compressive optic neuropathy
- Oil droplet cataracts
- Corneal pathology
Anterior Segment Pathology

- Absence of an afferent pupillary defect
- Oil droplet cataract: double density (SLE)
- Corneal warpage (keratoconus): corneoscope
- Corneal topography or Pentacam
- Retinoscope
DOM 25yo male

4/93: Bilateral visual loss while in jail

Va: 3/200 OU; no APD; Disc: normal
DOM – W/U
ERG: wnl
Serology:
VDRL: neg
ESR: nl
ANA: negative
CBC: nl
DOM - W/U

Leber’s genetic screen: + mutation at 3460

F/U (7mo): “Worse”

Va: 3/200 OU; N: 20/400 equiv OU
NFM 42yo female

11/92: 10mo h/o “blurred Va & trouble w/ colors”

Va: 20/70, 20/200; N: 6pt, 26pt; 3/10 HRR plates OU

VF:

Ext: w/q

P: w/o APD

EOM: full

SLE: wnl

Ta: 19/16

Fundus:
NFM – PMH

4 laparotomies w/ small bowel resection 1973-7

Rx: Parenteral multivitamins + hydroxycobalamin
NFM – F/U

12/92(1mo): “Better”

Va: 20/25 OU

N: 3pt, 4pt

VF:

Ext: w/q

P: w/o APD

SLE: wnl

Fundus:
NFM - F/U

7/93 (8mo): “Marked improvement”

Va: 20/20 OU

N: 3pt OU

VF:

Ext: w/q

P: w/o APD

SLE: wnl

Fundus: PM dropout
Commonly Missed Diagnoses

- Compressive optic neuropathy
- Oil droplet cataracts
- Corneal pathology
- Hereditary/Metabolic optic neuropathies
Hereditary/Metabolic Optic Neuropathy

- Central scotoma
- Discs may be normal early
- Ask for family history: maternal
- Previous GI surgery
BL 31yo male

5/04: Decrease vision

Va 20/100, 20/200

VF:

Ext: w/q

P: w/o APD

EOM: full

SLE: wnl

Ta: 15/14

Fundus:
BL – W/U

mERG:
Commonly Missed Diagnoses

- Compressive optic neuropathy
- Oil droplet cataracts
- Corneal pathology
- Hereditary/Metabolic optic neuropathies
- Maculopathy
Occult Maculopathies

- Metamorphopsia (Amsler grid)
- Lack of afferent pupillary defect (APD)
- Central scotoma w/o breakout
- OCT
- F/A, ICG
- mERG
RDH 36yo male

8/97: 2mo h/o vertical diplopia

Va: 20/15 OU; N: 3pt OU; no APD

Ext: no proptosis

AChR Ab +
Commonly Missed Diagnoses

- Compressive optic neuropathy
- Oil droplet cataracts
- Corneal pathology
- Hereditary/Metabolic optic neuropathies
- Maculopathy
- Myasthenia gravis
Myasthenia Gravis

- When pattern doesn’t fit cranial nerve (even when it does)
- When variable (worse when tired)
- Not associated with pain or pupillary changes
LAD 77yo female

5/96: Double vision “after cataract”

Va 20/20, 20/25; N 3pt, 4pt

VF: full

Ext: palp 9 OU, H 16 OU

P: w/o APD

EOM: RHT inc up gaze

SLE: PC-IOL

Ta: 20/19

Fundus: nl DMV
LAD – W/U

CT:
Commonly Missed Diagnoses

- Compressive optic neuropathy
- Oil droplet cataracts
- Corneal pathology
- Hereditary/Metabolic optic neuropathies
- Maculopathy
- Myasthenia gravis
- Thyroid orbitopathy
Thyroid Orbitopathy

- Not easy if no proptosis or prior hx thyroid disease
- Most common cause orbital pathology (50%)
- Imaging (enlarged EOM)
- Evidence of restriction (elevated IOP)
- Thyrotropin inhibitor binding assay
- Optic neuropathy only 5-8%
PAC 19yo male

4/84: MVA w/ “double vision”

CT: “normal”

Va: 20/15 OU; N: 3pt OU; VF: full

EOM: limitation adduction & abduction OS
PAC - Review of negative CT:

Look straight

Look left
PAC - W/U: IOP OS ↑ 17 → 30 w/ attempt abduction

Repeat CT:
Commonly Missed Diagnoses

- Compressive optic neuropathy
- Oil droplet cataracts
- Corneal pathology
- Hereditary/Metabolic optic neuropathies
- Maculopathy
- Myasthenia gravis
- Thyroid orbitopathy
- Other restrictive strabismus
Orbital Restriction

- Previous history of trauma
- Previous history of neoplastic disease
- Proptosis or other orbital signs
- Positive forced ductions or elevated IOP
- Imaging (CT) with coronals
HDM 61yo female

10/95: Acute blurred vision

“Nothing on eye exam”

Symptoms resolved in 4 days

Some associated headache

11/95: Double vision

CT: “normal”

MRI: “normal”

Dx: IVth nerve palsy
HDM - F/U

11/95: Pulsatile tinnitus

ENT: hearing normal

Rx: nortriptyline + Atenolol
HDM - F/U
1/96: Recurrent double vision
“Redness”
Dx: “infection”
Rx: Cipro

Progressive conjunctival prolapse
Refer to UVA
HDM - PE

Va 20/25, 20/400; N 3pt, 26pt

VF:

Ext: 3+ chemosis, H 22/24

P: .9-1.2 log LAPD

EOM: Absent abd OS, limit OD, 45ΔLET

SLE: wnl

Ta: 21/23

Fundus:
HDM - W/U

Angio:
Commonly Missed Diagnoses

- Compressive optic neuropathy
- Oil droplet cataracts
- Corneal pathology
- Hereditary/Metabolic optic neuropathies
- Maculopathy
- Myasthenia gravis
- Thyroid orbitopathy
- Other restrictive strabismus
- Carotid cavernous fistula
Carotid-Cavernous Fistula

- Episcleral venous engorgement not conjunctivitis
- Direct (high flow following trauma)
- Low flow (dural)
- Ask about bruit
- Look for increased pulse pressure
LGS 79yo male

4/03: Diplopia

Va 20/20, 20/80

N 3pt, 10pt

VF:

Ext: ptosis OS, H 16/20, ↓sens V1,2

P: .9log LAPD

EOM: complete ophthalmoplegia

SLE: 1+NS

Ta: 26/21

Fundus: nl DMV
LGS – PMH

2001: “Red spot” L temple

6/02: “Numbness L face”

8/02: Pain L face

Dental consult

Neurology: Neurontin

12/02: Worsen pain

MRI (1/03): “Periventricular white spots”

2/03: Neurosurgery consult for trigeminal neuralgia
LGS – POH

12/02: Oblique double vision

2/03: L ptosis
LGS – W/U

MRI:
LGS – W/U

MRI:
LGS – Rx
60Gy RT
Commonly Missed Diagnoses

- Compressive optic neuropathy
- Oil droplet cataracts
- Corneal pathology
- Hereditary/Metabolic optic neuropathies
- Maculopathy
- Myasthenia gravis
- Thyroid orbitopathy
- Other restrictive strabismus
- Carotid cavernous fistula
- Neurotrophic spread of cancer
Neurotrophic Spread

• Previous history of facial tumor (squamous)
• NUMBNESS – BAD
• When the imaging studies don’t fit the clinical finding recheck imaging
Anxiety Level

- Acute visual loss (especially normal disc)
- Visual field defects (especially if respect vertical)
- Acute painful ophthalmoplegia (especially if pupil involved)
- Numbness (with or without pain)
- Painful anisocoria
Mnemonics for All

- 5 A’s on a CD
  - Arteritis
  - Apex syndrome
  - Apoplexy
  - Aneurysm
  - Amaurosis

- Don’t forget C/D
  - Compression
  - Dissection
Conclusions

• Ocular malalignment (diplopia)
  – Restrictive
  – Paretic (not all CN): MG/skew

• All decreased vision not optic neuropathy
  – Anterior segment (lens; cornea)
  – Retina (maculopathy)
  – Importance of visual fields

• Orbital signs demand imaging

• Follow-up critical