Neuro-ophthalmic Diagnoses Not to Miss

Steven A. Newman, M.D.
Charlottesville, VA
Financial Disclosure

- Neuro-ophthalmology and financial interests: a contradiction in terms
- No I don’t have a law degree
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”
BAW – PMH

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
BAW – W/U
9/10: TA Bx:
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 - 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: >1.8log LAPD

EOM: full

SLE: 3+NS OD

Ta: 13 OU

Fundus:
CWA - W/U

OCT:
CWA - W/U

ESR: 17

CRP: 1.1
CWA - W/U

TA Bx: GCA
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)
CHH 71yo male

2/97: Sudden bilateral loss Va

Va: NLP OU

2wks before Va: 20/40, 20/200; P: “miotic”; Ta: 18 OU
W/U:
CT:
MRI:
Rx: TSS
No return Va
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
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):
PE – Rx

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
Severe Consequences

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

- Pupil sparing not present
- Potential for missing w/ MRA/CTA
- Sentinel bleeds
SFR 59yo female

9/99: 2-3mo h/o ↓Va OD X 1-2min

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

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
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

ODM: 90/45; 105/50
NFL – W/

Cocaine test:
NFL – W/U

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 vs spontaneous
- Risk: fibromuscular dysplasia, Ehlers-Danlos IV
- Symptoms:
  - Facial pain
  - Horner’s syndrome (58%)
  - Dysgeusia
- Consequence
  - Hemispheric stroke
  - CRAO/BRAO
AWB 69yo male

3/83: Decreased Va → Dx: “macular degeneration”

Referred for laser but cancelled after negative F/A

5/84: ECCE OS → vision no better, Dx: PC opacification

3/87: YAG OS → vision no better

9/87: ↓ Va OS → Dx: RD OS; Rx: SBP OS

No return Va
AWB – F/U
11/87: Referred to UVA
Va: 20/25, 1/200
>3log LAPD
AWB - W/U:
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
Anxiety Level

- Acute visual loss (especially normal disc)
- Visual field defects (especially if bitemporal)
- 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

- Elderly patient = GCA
- VF testing across vertical midline EVERY EXAM
- Motility abnormality + ↓Va implies orbital apex requires imaging
- Pupil involved IIIrd = aneurysm until excluded
- TMO requires evaluation for thromboembolism
- Horner’s implies dissection until excluded
- FOLLOW-UP is CRITICAL