Case 1

25 year old woman

- 1.5 years ago – routine exam uncovered increased cupping (OS > OD)
- 3 yrs ago, C/D was 0.3 x 0.3 OD & OS
- Due to increased C/D, optometrist spoke with PCP, and PCP ordered MRI of brain and orbits with and without orbits
- MRI reported to be normal

RAPD GRADING

- 1+ = Early Release / Escape
- 2+ = No Initial Movement Followed By Early Release
- 3+ = Immediate Release
- 4+ = Amaurotic Pupil - NLP

Brightness Sense Comparison

Appearance of Bright Light with Decreasing Optic Nerve Function
Mean deviation OS: -12.25 DB
Mean deviation OD: -0.25 DB
Difference: 12.00 DB
Divide by 10: 1.2 DB
(1.2 log-unit NDF needed OD to balance RAPD OS)

Significant RAPD OS evident on pupil testing.

An RAPD can ONLY occur with lesions at an anterior to the Lateral Geniculate Nucleus (LGN)!

5 NEURO-OPHTHALMIC DIAGNOSTIC COMPARISONS

- VISUAL ACUITY to COLOR VISION
- VISUAL FIELD to RAPD (visa versa)
- RAPD to OPTIC DISC
- OPTIC DISC to VISUAL FIELD (visa versa)
- VISUAL ACUITY to VISUAL FIELD

HVF has shown only mild progression since then
Pt is now noticing a blurry spot in OS
Denies any new symptoms
Pt no longer has insurance
- VA OD 20/20 OS 20/30
- Color OD 14/14 OS 9/14
- PERRL (+ 0.9 log RAPD OS)
- CF: central temporal red desaturation OS

- VF respects the vertical meridian
- Suspect suprasellar lesion
- Ask pt to obtain previous MRI films

Case 2
77 year old man

- Reports 3 week history of blurred vision OD
  - Notices especially when reading
  - Right-sided weakness
- Visual acuities 20/20 OD 20/20 OS
- PERRL (trace +) RAPD OD
- Confrontation fields: right homonymous hemianopia denser superiorly
- Medical history
  - Hypertension

Visual Field Results

Medical History

- Hypertension

Visual Field Results

Lesion of left optic tract

Nasal and papillomacular fibers cross in the chiasm

Incongruous right homonymous hemianopia

Bow-tie (band) optic atrophy

- Optic tract lesion
- Ipsilateral ST/IT pallor
- Contralateral band pallor (temporal VF defect)
  - From nasal macular fibers (papillomacular bundle)
  - May have small RAPD in contralateral eye
  - Incongruous homonymous hemianopia
Case 3

50 year old woman

- Vision loss OU gradually x 4 months
- Decreased color vision
- Sees flashing lights
- No headaches or eye pain
- Hearing loss x years
- No other reported neurologic symptoms
- (+) HTN, hypercholesterolemia
- Meds: HCTZ (thiazide diuretic), lisinopril (ACE inhibitor), sular (Ca channel blocker)
- (-) significant past medical history

Previous Eye Exams (records obtained)

- 6 months prior (at an outside facility)
  - VA OD 20/30 OS 20/30
  - Pupils poorly reactive
  - Further evaluation requested

- 3 months prior (at a different outside facility)
  - VA OD 20/200 OS 20/80
  - Mild pallor noted
  - MRI ordered (reported to be normal)
  - Neuro-Ophthalmology consult requested

Neuro-Ophthalmology Evaluation (outside facility)

- Severely constricted VF's by Goldmann
  - 5 to 7 degrees in each eye
- VA OD CF@3ft OS CF@3ft
- Diagnosed with functional vision loss
- No additional work-up requested
- Follow-up scheduled in 6 months

Pt presents for another opinion

- BCVA OD 1/600 OS 10/500
- Normal ocular motility exam
**LIGHT/NEAR DISASSOCIATION PUPILS**
(5 Causes)

- AMAUROTIC (blind eye)
- TONIC
- ARGYLL ROBERTSON
- TECTAL (Dorsal Midbrain Syndrome)
- ABERRANT REGENERATION OF CN III

**IS THIS AN ARGYLL-ROBERTSON PUPIL?**

- MIOSIS (2.5 mm in dark)
- ABSENT DIRECT RESPONSE TO LIGHT
- BRISK NEAR (LND)
- PRESERVED VISION
- UNILATERAL, ASYMMETRIC OR UNEQUAL
- DILATES POORLY
Work-up

• MRI reported to be normal
• Lab Testing remarkable for:
  – Reactive FTA-ABS
  – Reactive RPR 1:8 titer
  – Low folate level (3)
  – Elevated homocysteine level (21)

Treatment

• Pt admitted to hospital for LP
  – (+) VDRL
  – Diagnosis of NEURO-SYPHILIS
  – Always check for co-existent HIV
• Initiation of IV Penicillin x 14 days

• Low vision services

Case 4

52 year-old man

• Family indicates he lost 20 lbs in past yr
• Problems with walking and balance
• He keeps holding his chin up
• Changes in mental status and behavior
  – Pt thinks all problems are from glasses & clothing
• Hasn’t seen a doctor in > 10 years

• VA: OD 20/40 and OS 20/40
• Color: OD 7/7 and OS 7/7
• CF: full OU
• Palpebral apertures: OD 10 mm and OS 10 mm
• Exophthalmometry: OD 17 mm and OS 17 mm
• Normal SLE
• TA: OD 14 mm Hg and OS 17 mm Hg
• DFE: normal optic nerves and retina OU
• Neurologic exam:
  – Broad-based, ataxic gait
  – Positive Rhomberg sign
  – Slow, slurred speech
DORSAL MIDBRAIN SYNDROME

- TECTAL PUPILS
- UPGAZE PARESIS
  (DOWNGAZE PARESIS, OR BOTH)
- CONVERGENCE RETRACTION NYSTAGMUS
- EYELID RETRACTION
Diagnosis

- Multiple lesions noted, not only in midbrain, but throughout brain
- Characteristic of metastatic lesions
- No known history of a primary cancer
- Work-up to find primary site revealed multiple organs involved

Etiology of DMS

- Compression of dorsal, rostral midbrain in region of posterior commissure
  - Tumor
  - CSF obstruction
  - Inflammatory
  - Infection
  - Metastatic

Work-up

Start with Neuro-imaging

Case 5
Clinical Features

- “Flat” edges
- “Vermiform” iris movement
- Poor response to light & near or LND
- “Dilation lag” following prolonged near effort
- “Paradoxical Pupil” - aniso greater in light & dim
### Pathogenesis of Adie’s Tonic Pupil

- Ciliary ganglion
  - 90% CB
  - 3% iris
- Aberrant regeneration of CB fibers to iris sphincter (light-near/gaze pupil dissociation)

Adie WJ. Brain 1932

### Local Tonic Pupil
- Varicella
- Retrolubar
- Orbital Tumor
- Orbital Surgery

### Neuropathic Tonic Pupil
- Diabetes
- Syphilis
- Sarcoid
- Lyme

### Idiopathic Tonic Pupil
- “Adie’s”
- Unknown Etiology

### Case 6

**Initial Presentation**
Right Horner syndrome suspect due to right-sided ptosis and miosis.

### Diagnostic Test For Horner Syndrome

- 0.5% or 1.0% Apraclonidine (Iopidine)
- Alpha agonist
- Weak alpha 1 agonist
- No effect on normal pupil
- Dilates Horner pupil (supersensitivity)
- Look for **REVERSAL OF ANISOCORIA**
- May NOT be positive in acute Horner Syndrome

### Normal Eye

- Norepinephrine production controlled by alpha 2 receptors, which work by down-regulation of alpha 1 receptors responsible for dilation

**Horner Eye**

- Norepinephrine amount is reduced, so alpha 2 receptors are not activated, resulting in up-regulation of alpha 1 receptors, leading to denervation sensitivity
Initial Presentation

Right Horner syndrome suspect due to right-sided ptosis and miosis.

Case #1

Initial Presentation

Digital infra-red photos taken under scotopic illumination. Note reversal of anisocoria after use of Apraclonidine, indicative of a right Horner syndrome.

Post-Apraclonidine

Demonstrates ease of detection of reversal of anisocoria, indicating a positive test for Horner syndrome.

Case #2

Initial Presentation

Horner syndrome suspect due to left-sided ptosis and miosis.

Post-Apraclonidine

Negative result – no reversal of anisocoria after use of Apraclonidine.

Case 7
• 42 yo healthy man watching TV with neck flexed x 2hr.
• Jumps up for phone call
• **Within 1 hr.** recurrent “black spot”, OD
• **Within 2 hrs.** ipsilateral exploding headache, right eyelid droop
• **Next 12 hrs.** right jaw pain, dysguesia

A **PAINFUL** Horner Syndrome is a Carotid Dissection Until Proven Otherwise!
**Carotid Artery Dissection**

- Need to consider this diagnosis in EVERY **PAINFUL HORNER's**
- Can occur with or without trauma
- Medical Emergency
- Horner's with eye, head, neck pain
- Pt to hospital (MRI, MRA, CTA, angiogram)

**Diagnostic Test For Horner Syndrome**

- 0.5% or 1.0% Apraclonidine (Iopidine)
- Alpha agonist
- Weak alpha 1 agonist
- No effect on normal pupil
- Dilates Horner pupil (supersensitivity)
- Look for REVERSAL OF ANISOCORIA
- May NOT be positive in acute Horner Syndrome
  - Such as in carotid dissection

**Case 8**
**68 year-old man**

- Diplopia x 4 days
- Undergoing chemo (Rituxan) for non-Hodgkin’s Lymphoma
- D/C chemo due to diplopia
- Had MRI of brain without contrast (no etiology for diplopia)
- Constant frontal headache x 7 months
- Pain worse x 4 days – over and behind right eye
- Has been closing right eye to avoid diplopia

- + DM x 10 yrs, HTN since age 18
- + hypercholesterolemia
- S/p MI x 2
- S/p CABG X 4
- + Atrial Fibrillation (on Coumadin)
- S/p Parathyroidectomy
- MEDS: Glucotrol, Coreg, Digoxin, Pravachol, Zoloft, Synthroid, Coumadin, amitriptyline, Protonix, Colchicine

**PUPIL IN CN III Palsy**

- INVOLVED = ANEURYSM (86%)
- SPARED = VASCULOPATHIC (77%)

**DOES NOT APPLY IF:**
- COMPLICATED CN III
- INCOMPLETE CN III
- RELATIVE SPARING
- 20-50 YEARS OF AGE

**PAIN in CN III Palsy**

- ANEURYSM = 95%+
- DIABETES = 80%

PITUITARY APoplexy, GIANT CELL, CAVERNOUS SINUS

**CN III PALSY WORK-UP**

**ADULTS**

- **20-50 YEARS**
  - CT, MRI, MRA, A-GRAM

**CHILDREN**

- CONGENITAL (MRI)

**ACQUIRED**

- EXCLUDE TRAUMA OR MIGRAINE
- CONSIDER LP IF MRI (-)
- IF MRI & LP ARE NEGATIVE > 10 years, ARTERIOGRAM TO LOOK FOR ANEURYSM

**COMPLETE CN III – PALSY**

- Can use the pupil as a guide

**INCOMPLETE CN III – PARESIS**

- Can NOT use the pupil as a guide

**20-50 YEARS**

- CT, MRI, MRA, A-GRAM

**50+ YEARS**

- PUPIL, PALSY, PAIN
- NEUROIMAGING
- VASCULOPATHIC EVALUATION
- R/O GCA
1 month later

48 year old woman

HA and pain OD x 2 weeks

Went to ER

Initially told symptoms were from the flu

VASCULOPATHIC

ANEURYSM

EMERGENCY: Sub-Arachnoid Heme 20% die in 48 hrs!

PSEUDO – GRAEFE SIGN

LIGHT-NEAR DISSOCIATION PUPILS

ABERRANT REGENERATION OF CN III

Aneurysm, Tumor, Trauma

NEVER Diabetes!

EYELID SYNKINESIA