DIAGNOSIS AND MANAGEMENT OF NEURO-OPHTHALMIC DISEASES: RULES, EXCEPTIONS TO THE RULES, AND EXCEPTIONS TO THE EXCEPTIONS TO THE RULES

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Thurston Howell III Doesn’t Like Neuro

“Neuro equals referral”

“Diagnose and adios!’
Managing Patients with Neuro-ophthalmic Disease

- Understanding of anatomy
- Following several fundamental principles
- Following several simple rules
- Developing a network of referral physicians
  - Neuroradiologist
  - Neurologist
  - Internist
  - Neurosurgeon
  - Rheumatologist
A personal case to prove my point
A Personal Case to Prove My Point
Bitemporal visual field defects mimicking chiasmal compression in eyes with tilted disc syndrome

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Abstract

BACKGROUND: Tilted disc syndrome (TDS) is a congenital optic nerve coloboma occurring from embryonic dysgenesis. Several features characterize TDS, including an inferiorly located conus, situs inversus of the major retinal vessels, and an anomalous disc shape. Commensurate with axonal dysgenesis, visual field defects may often occur from TDS, the most common of which involve the temporal and superior temporal visual field. These visual field defects can mimic those seen in chiasmal compression from a mass lesion.

CASES: Five patients from New South Wales, Australia, with distinct TDS and bitemporal visual field defects on frequency doubling threshold perimetry seemingly respecting the vertical hemianopic line are presented. Neuroimaging and medical evaluation of each failed to show intracranial chiasmal pathology in any patient.

CONCLUSIONS: TDS can present with visual field loss resembling that seen in chiasmal disease. Although most cases of temporal visual field loss from TDS do not respect the vertical hemianopic line and are not true quadrantanopsias, there are instances in which this does occur, likely caused by the testing modality used. It is essential that patients with suspected intracranial pathology undergo immediate neuroimaging, even in the face of TDS.

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Tilted disc syndrome (TDS) is a congenital defect of the optic nerve. Although the appearance may vary among inferior nasally where the choroidal vessels are more readily visible. Frequently, there is situs inversus of the major retinal
You’re wife is going to kill you if she finds out!
Rule

Congenital optic nerve anomalies can have (sometimes dramatic) visual field loss
A (insert any age) (insert either sex) patient with a previous history of cancer presents with (insert any neuro-ophthalmic finding). The patient also complains of (insert any symptom). Additionally, you note (insert any finding). What is the most likely cause?

a. Cancer
b. Cancer
c. Cancer
d. All of the above
Never diagnose idiopathic anything in a patient with a history of cancer
Don’t make diagnosis of immune disease in immunosuppressed patients
Rules Must be Obeyed

- 57 YOF
- Low risk OHTN OU
- GDx, OCT, ONH – perfectly normal OU
Fields are a different story however...
So, where is the lesion?

So, where is the lesion now?

Apparently, nowhere
Chiasmal and retrochiasmal lesions have bilateral involvement.

Unilateral visual field loss reflects anterior visual pathway disease which will show something identifiable in the form of damage to the vision disc, RNFL, dyschromatopsia or afferent pupil defect.
A patient can fake a field, but can’t fake a retinal nerve fiber layer or pupil defect.
59 YOM

- Routine exam - c/d 0.5/0.5 OU
  - IOP 20 mm Hg OU
- Returns 2 years later - slowly progressive loss of vision OD
- RAPD OD; 20/80 OD; 20/20 OS
- Superior altitudinal defect splitting fixation OD; mild inferior defect OS
- Disc pallor OD
- Dx: NAAION

What is wrong with this picture?
59 YOM

- Routine exam- *c/d 0.5/0.5 OU*
  - IOP 20 mm Hg OU
- Returns 2 years later- slowly progressive loss of vision OD
- RAPD OD; 20/80 OD; 20/20 OS
- **Superior altitudinal defect** splitting fixation OD; mild inferior defect OS
- Disc pallor OD
- Dx: NAAION

What is wrong with this picture?
59 YOM

- IOP 23 mm Hg OD
- c/d actually 0.95/0.95 OD and 0.8/0.8 OS
  - Very shallow cupping
- Dx: undiagnosed POAG with loss of fixation OD
Rule

Don’t make the diagnosis of NAAION in glaucoma patients
Rule

A diagnosis of exclusion (Adies tonic pupil, PTC, Bell’s palsy, NAAION, Tolosa Hunt syndrome) should your last diagnosis, not your first
48 YOWM

- Painless loss of visual field OS
  - 20/20 OD, OS
  - Noticed upon waking
- Med Hx: Unremarkable, except for viral illness 3 weeks before
NAAION OD
Disc at risk OS
Rule

Pallor in excess of cupping indicates something other than, or in addition to, glaucoma.
Nothing notches a nerve like glaucoma
In the Age of Imaging, Do We Really Need Fields?

- 54 YO Nigerian man
- Referred for glaucoma management
- Told he had glaucoma 6 years earlier- no Tx
- 20/30 OD; HM OS
  - Vision loss from glaucoma- not coming back
- 30 mm Hg OD; 23 mm Hg OS
  - Lumigan- 17 mm Hg OD, 15 mm Hg OS
Diagnosis?

Plan?

Do we really need fields in this case?
Yes, we still need to do fields in the age of imaging.

Sometimes its not glaucoma
Ode To a Cupped Disc

Oh, to have a cupped disc pink.
That my friend hath a glaucomatous stink.
But to have a cupped disc pale,
Call this glaucoma and you shall fail.
Disc and field damage that is one-sided
Simply cannot be abided.
It might be trauma, infarct or meningioma.
But if the rim is cut always remember,
Nothing notches a nerve like glaucoma.
POAG gets complicated?

- 70 YOWM
- POAG OU
- Auto accident with concussion
- Develops gaze induced amaurosis fugax
- Referred by PCP to neuro-ophthalmologist
- Complete evaluation with MRI- negative
- Psychological?
Sometimes it is glaucoma
Case History 46 WM

- CC: Patient reports a "droopy left eye" which began about 6 weeks ago. Headache and numbness ipsilateral; hives
  - ER diagnosed with "stye". Patient was referred in by a local optometrist.
- Past Ocular History: unremarkable
- Past Medical History: (+) Mitral Valve Prolapse, (+) GERD and recent weight loss of about 20 lbs. over the past 6 months or so.
  - Medications: Prilosec, Metoprolol Succinate, Xanax, Prednisone, Lipitor, Claritin
Pertinent Findings

- BCVA 20/20 OD and 20/20 OS
- Pupils: *unequal*, round, reactive to light, No APD

<table>
<thead>
<tr>
<th>Bright Illumination</th>
<th>Dim Illumination</th>
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</thead>
<tbody>
<tr>
<td>OD: 4 mm</td>
<td>OD: 6 mm</td>
</tr>
<tr>
<td>OS: 3 mm</td>
<td>OS: 4 mm</td>
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</tbody>
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- Motility and confrontation fields unremarkable
- Observation: LUL ptosis, Left miosis
- Intraocular pressure: 18 mmHg OD and 19 mmHg OS
- Fundoscopy-unremarkable
So, what do you think and what do you want to do now?
Post-Iopidine
Horner’s Syndrome

- Etiology unclear based upon exam
- Headache, neuralgia and ‘hives’
  - Not consistent with cluster migraine
    - Dx of exclusion, not convenience
  - Hives- not consistent with HZO
- Unexplained weight loss concerning- relationship unclear
- Recommend medical eval by PCP
  - Additional testing dictated by PCP results
Discussion

- What is Horner’s Syndrome?
  - a triad of clinical signs arising from disruption of sympathetic innervation to the eye and ipsilateral face that causes *miosis*, upper lid *ptosis*, mild elevation of the lower lid, and *anhydrosis* of the facial skin.
Pharmacological Testing

- **Cocaine**
  - Horner’s pupil doesn’t dilate, normal pupil does

- **Hydroxyamphetamine (Paredrine)**
  - Differentiates post- from pre-ganglionic
  - Not available and doesn’t matter because bad stuff happens everywhere

- **Apraclonidine 0.5% (Iopidine)**
  - Denervation suprasensitivity
  - Horner’s pupil dilates, normal doesn’t
    - Reversal more classic and diagnostic than cocaine
Horner’s Syndrome: Etiologies

- First-order neuron disorder: Stroke (e.g., vertebrobasilar artery insufficiency or infarct); tumor; multiple sclerosis (MS), and, rarely, severe osteoarthritis of the neck with bony spurs.

- Second-order neuron disorder: Tumor (e.g., lung carcinoma, metastasis, thyroid adenoma, neurofibroma). Patients with pain in the arm or scapular region should be suspected of having a Pancoast tumor. In children, consider neuroblastoma, lymphoma, or metastasis.
Horner’s Syndrome: Etiologies

- Third-order neuron disorder: Headache syndrome (e.g., cluster, migraine, Raeder paratrigeminal syndrome), internal carotid dissection, herpes zoster virus, otitis media, Tolosa–Hunt syndrome, neck trauma/tumor/inflammation, prolactinoma.
- Congenital Horner syndrome: Trauma (e.g., during delivery).
  - Facebook tomography
- Other rare causes: Cervical paraganglioma, ectopic cervical thymus
Treatment and Management

- Localizable- targeted workup
  - Neck and facial pain- carotid dissection
  - Facial paraesthesia- middle cranial fossa disease

- Necessary Work Up (non-localizable):
  - MRI of brain, orbits and chiasm with and without contrast, attention to middle cranial fossa.
  - MRA of head and neck-rule out carotid dissection
  - MRI of neck and cervical spine, include lung apex and brachial plexus
    - Horner’s syndrome patient needs to be imaged from chest to head- 3 scans
    - Horner’s protocol

- All imaging in patient unremarkable
Carotid Dissection

- A 3rd-order Horner’s and ipsilateral head, eye, or neck pain of acute onset should be considered diagnostic of internal carotid dissection unless proven otherwise.
Carotid Dissection

- Carotid artery dissection presents with the sudden or gradual onset of ipsilateral neck or hemicranial pain, including eye or face pain.
- Often associated with other neurologic findings including an ipsilateral Horner’s syndrome, TIA, stroke, anterior ischemic optic neuropathy, subarachnoid hemorrhage, or lower cranial nerve palsies.
- Horner’s from suspected carotid dissection should go to ER.
Rule

Diagnosing Horner’s syndrome is insufficient. You must try to ascertain a cause and never assume that it is benign.
Case: 59 BF

- Long time patient presents for her glaucoma f/u. She reports drooping in the right eye and smaller pupil for about 1 month. Symptoms were noticed at/ about time of dx of lung cancer and subsequent surgery.
  - She also reports scapular pain and weakness in the right hand.
- Past Medical History: (+) Lung Cancer, (+) Pancreatitis, (+) HTN and (+) Acid Reflux
- Social History: Smokes 1 pack per day for 45 years, Drinks a 6 pack of beer daily
Case 2: Pertinent Findings
Continued...

- Pharmacological testing not done
- New onset of ptosis and miosis with dx lung cancer and h/o recent lung surgery
- Dx=Pancoast Syndrome
Pancoast Tumor

- A Pancoast tumor is a lung cancer arising in the apex of the lung that involves structures of the apical chest wall.

Treatment

- Chemotherapy
- Radiation Therapy
- Surgery: lobectomy vs. wedge resection

Prognosis: 5 year survival rate is around 30%
Case:

23 Year Old White Female

- **CC:** Sudden onset pupil dilation with ipsilateral headache
- **Medical Hx:** normal
- **BVA:** 20/20 OD, OS
- **Pupils:**
  - 3 mm anisocoria, OS larger, anisocoria greater in bright illumination. Previously isocoric. (-) RAPD, (+) Accom
- Remainder of exam normal
- Similar incident 2 days antecedent, resolved within hours
- What does she look like?
Case:
23 Year Old White Female

What questions do you want to ask?

What tests do you want to order?
Case:
23 Year Old White Female

Additional questions to ask:
- **Any double vision?** No!
- **Any use of ophthalmic pharmaceuticals?** No!
- **Any history of migraine headaches?** Maybe...

**Differential diagnosis?**
- Aneurysmal compression on CN III? No!
- Pharmacological misadventure? No!
Benign Episodic Pupillary Mydriasis

- Episodic unilateral mydriasis
  - Lasts minutes to weeks
- Accompanied by blurred vision and headache
- Young, healthy females (*may have migraine history*)
- Peculiar sensations about affected eye
  - Often progresses to headache
  - Not typical migraine
- Defective accommodation
- Lid and motility defects *not* present
- Extensive medical testing *unremarkable*
Benign Episodic Pupillary Mydriasis

- Increased sympathetic activity?
  - Reverse Horner’s syndrome – not likely

- Pupil paralysis following migraine?
  - Tends to last longer – not likely
  - No ophthalmoplegia

- Spasm of segment(s) of iris dilator muscle?
  - Round pupil, so not likely

- Pharmacologically dilated?
  - Parasympatholytic – no light or near reactivity
  - Sympathomimetic – can mimic and must R/O
Benign Episodic Pupillary Mydriasis

- Anisocoria greater in bright than dim
  - Parasympathetic dysfunction
    - Not an aneurysm
    - Edinger-Westphall lesion?
- Migraine variant – most likely etiology
- Treatment – none except to avoid unnecessary testing
Pupil Rules

- Anisocoria greater in dim = sympathetic dysfunction
  - Horner’s syndrome - look for dilation lag
  - Miotic use
- Anisocoria greater in light = parasympathetic dysfunction
  - CN 3 palsy
  - Tonic pupil
  - Pharmacologic or traumatic pupil
    - No reactivity?
Pupil Rules

- Fixed and dilated and unresponsive to light or near = pharmacologic or iris trauma
Rule: **Isolated Dilated Pupil Is Almost NEVER An Aneurysm**

Ambulatory patients with isolated dilated pupil more likely to harbor iris or ganglion (Adie’s) lesion or medication misadventure than CN 3 palsy

Comatose patient is a different story

Risk of angiography is much higher than risk of aneurysm in this setting

No imaging needed for isolated dilated pupil
Rumors about Bowie’s Pupils

- Dilated from injecting heroin
- Attacked by a gang in hate crime
- Reality: Traumatic anisocoria at age 13 years in fight with best friend George Underwood over a girl!
- Permanently dilated pupil
- Hazel with rim of blue
Rule

Don’t neuroimage David Bowie
47 Year Female

- **CC:** Horizontal double vision in far left gaze
- **BVA:** 20/20 OD, OS
- **Medical Hx:** newly diagnosed diabetes
- **Left abduction deficit in far left gaze**
  - Negative forced duction test
- **Mild ocular injection OS**
- **IOP:** 14 mm Hg OD, 16 mm Hg OS
- **Fundus:** normal OU

Thoughts?
47 Year Old Black Female

- **Presumptive diagnosis:** Left vasculogenic CN VI palsy - monitor
- Returns 1 week with marked worsening of injection, diplopia and ophthalmoplegia
- **IOP:** 16 mm Hg, 26 mm Hg
- Fundus disc congestion and vascular tortuosity OS

**What does she look like NOW?**

**What do you want to do NOW?**
47 Year Old Black Female

- CT scan:

What do you think NOW?
Carotid Cavernous Sinus Fistula

- **Cavernous sinus**...
  - Trabeculated venous cavern
  - Houses CN III, IV, VI, V1, oculosympathetics, and ICA
  - Drains eye and Adnexa via inferior and superior ophthalmic veins to petrosal sinuses and jugular vein

- **Fistula**...
  - Rupture of ICA or meningeal branches within sinus
    - Meningeohypohyseal, McConnell’s Capsular, Inferior Cavernous
  - Mixing of arterial blood in venous system
Carotid Cavernous Sinus Fistula

- Hemodynamic
  - High flow vs low flow

- Angiographic
  - ICA vs meningeal branches

- Etiology
  - spontaneous vs traumatic
Carotid Cavernous Sinus Fistula

- Increased venous pressure
- Orbital congestion
- Proptosis (pulsatile)
- Corneal exposure
- Arteriolization
- Orbital bruit
- Myopathies and cranial neuropathies with diplopia
- Secondary glaucoma
Carotid Cavernous Sinus Fistula

- Vision threatening – not life threatening
- Spontaneous etiology – spontaneous resolution
  - ICA compression with contralateral hand
- Traumatic – clipping and ligation
- Balloon or particulate embolization
- Manage glaucoma aggressively
  - prostaglandin analogs
Rule: Beware the Chronic Red Eye

- Dilated & tortuous episcleral vessels that go to the limbus and back (omega loops) π
- Intervening “clear conjunctiva”
- Red eye that doesn’t respond to any topical treatments
  - Bag-o-Meds
- Other non-red eye findings: Chemosis, IOP elevation, proptosis, ophthalmoplegia, ptosis, lid edema