Neuro-Ophthalmic Cases That Gave Me a Headache

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Why OD’s are Perfectly Positioned
• Several cranial n. associated with sensory or motor to eye, adnexae
• OD’s may be the first health care provider to encounter patient
• In many cases, time is critical!
• OD’s uniquely positioned to detect and evaluate conditions early

Cranial Nerves: Review
• Cranial nerves leave from brain, not spinal cord
• May be sensory, motor, or both
• Neuro-ophthalmic conditions rarely occur in isolation (but may on occasion)
• Understanding anatomy critical to reaching correct diagnosis
### Cranial Nerves

<table>
<thead>
<tr>
<th>#</th>
<th>name</th>
<th>sensory motor</th>
<th>nucleus</th>
<th>function</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Olfactory</td>
<td>S</td>
<td>Olfactory</td>
<td>smell</td>
</tr>
<tr>
<td>II</td>
<td>Optic</td>
<td>S</td>
<td>Ganglion cells retina</td>
<td>innervates: levator superior rectus</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>superior rectus inferior rectus inferior oblique</td>
</tr>
<tr>
<td>III</td>
<td>Oculomotor</td>
<td>1* M</td>
<td>Oculomotor</td>
<td>innervates:levator superior rectus</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>superior rectus inferior rectus inferior oblique</td>
</tr>
<tr>
<td>IV</td>
<td>Trochlear</td>
<td>2* M</td>
<td>Trochlear</td>
<td>Superior oblique</td>
</tr>
</tbody>
</table>

S= sensory, M = motor, S + M mixed*

### Cranial Nerves

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<th>function</th>
</tr>
</thead>
<tbody>
<tr>
<td>V</td>
<td>Trigeminal</td>
<td>S + M</td>
<td>Trigeminal Sens, spinal, motor</td>
<td>Sensory form face, muscles of mastication</td>
</tr>
<tr>
<td>VI</td>
<td>Abducens</td>
<td>M</td>
<td>Abducens</td>
<td>Abduct eye</td>
</tr>
<tr>
<td>VII</td>
<td>Facial</td>
<td>S + M</td>
<td>Facial, Solitary, Sup. Salivary</td>
<td>Sensory- taste Motor- lacrimal gland, salivary gland, facial expression</td>
</tr>
<tr>
<td>VIII</td>
<td>Vestibulo-cochlear</td>
<td>S</td>
<td>Vestibular Cochlear</td>
<td>sound, rotation and gravity</td>
</tr>
</tbody>
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S= sensory, M = motor, S + M mixed*
Case 1: I think I had a stroke!!!

- An 18 year old male presents complaining of a fixed, dilated left pupil—his mother is concerned about stroke
- He denies previous anisometropia
- Salient history included trauma to the left eye and the surrounding orbital tissue
- He is taking an unidentified OCT eye drop.
- He also has a history of working on a farm
Case 1: I think I had a stroke!!!

- General health history
  - No personal Hx of
    - DM, HTN, autoimmune disease
  - Denies
    - Alcohol use, smoking, recreation drug use

Clinical Findings

- Presenting visual acuities were
  OD 20/20  OS 20/25
- Pupils; OD 3 mm OS 7 mm
  OD brisk constriction,
  OS trace constriction
- EOMs, no restriction in any position
- Fields full to finger counting confrontation

What is your next move?

1. Send him home to sleep it off
2. Order CT scan of brain, orbits
3. Dilute pilocarpine test
4. Cocaine test
5. (RPR) rapid plasma reagin
   or(VDRL) venereal disease research laboratories test

See discussion at end of handout
Case 2: I see two; It hurts to chew, and I am tired too!

- A 72 year old retired male nurse presents with sudden onset diplopia. He also complains of fatigue. He has been hypertensive for many years and reports good control. He has undergone cataract surgery OU and enjoys good vision.

Case 2

- VA: 20/20 OU
- Cover test- 6 Δ esotropia which increases with L gaze- absent in R gaze
- Pupils: OD 3 mm OS 3.5 mm no APD
- BP 119/66 mm Hg
- SLE: pseudo with clear capsulotomy

Case 2

What do you think is wrong with him?

What would you do now?

1. Refer for CT scan
2. Refer for neuro consult
3. Order MRI of brain and orbits
4. Order sed rate & C-reactive protein
Case 2

One month later

• PCP ordered a heart cath-pending
• Patient now c/o
  – Diplopia greater w/ L gaze
  – Drooping of the right eyelid
• He also reports problems with chewing

Case 2

• VA 20/20 OU
• Pupils: OD 3 mm OS 3.5 mm no APD
• Δ esotropia which increases with L gaze- absent in R gaze
• 2 mm ptosis OS

Case 2

What is your diagnosis?
1. Cavernous sinus fistula
2. Temporal arteritis
3. Myasthenia gravis
4. Pituitary adenoma
5. Two of the above
**Case 2**

What would you do now?

1. Refer for CT scan
2. Refer for neuro consult
3. Order MRI of brain and orbits
4. Order sed rate & C-reactive protein
5. Refer to mortician

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**Case 2**

What is your diagnosis?

- Cavernous sinus fistula
- Temporal arteritis
- Myasthenia gravis
- Pituitary adenoma
- Two of the above

See discussion at end of handout

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**Case 3: First herpes, now this!**

- A 66 year old female paralegal presents with a complaint of recent onset anisocoria. One month prior to this visit, she was successfully treated for herpes simplex keratitis with gancyclovir ophthalmic gel (Zirgan) and systemic acyclovir. She is concerned RE the possibility of a CNS lesion.
Case 3: First herpes, now this!

- The patient reports very high stress levels; her only daughter got married a week after the HSK presented.
- Her general health is “excellent”; she has a history of osteoporosis and hypothyroidism.
- Medications include Avista and thyroid supplement.

Case 3: First herpes, now this!

- VA OD 20/20 OS 20/30
- BP 149/85 mm Hg
- SLE
  - Well centered SiHy CL, small scar OS in area of HSK lesions (cleared), slight area of reduced pigment at pupil margin OS
  - Pupils; OD 3 mm OS 5 mm fixed

Case 3: First herpes, now this!

- Tests
  - VF nl OU
  - EOMs nl OU
  - Dilute pilocarpine test- no constriction
  - Cotton wisp test:
    • OD nl sensation
    • OS reduced sensation
Case 3: First herpes, now this!

What is your diagnosis or next move?
1. Adies tonic pupil
2. Argyll-Robertson pupil
3. Order MRI orbits & brain
4. Herpetic sphincter atrophy
5. Post wedding stress syndrome

Case 4- A tale of two fools

• A 65 year-old female presents complaining of ocular dryness, reduced vision which is especially apparent at night. Her history is significant for longstanding dry eye associated with hypothyroidism and long-term use of a beta-blocker for hypertension

Case 4

• VA with refraction
  – OU 20/20
• Pupils 3/3 no APD
• EOMs unrestricted
• BP 137/74 mmHg
• SLE- SPK, gr II+ NS OU,
• FDT- see following slide
Case 4

Based on these findings what is our next move
1. Rx glasses, ignore FDT
2. Order MRI
3. Order threshold visual fields
4. Consult PCP
5. Order GDX

Case 4

Based on history & these threshold visual fields, what is your diagnosis
1. Primary open-angle glaucoma
2. Compressive optic neuropathy
3. Poor quality visual fields
4. Hysterical field loss
5. Temporal arteritis
Neuro report

“I think it is most likely that this represents asymptomatic residual of her history of Graves' disease. Her neurological examination is normal. I think it would be most prudent to simply repeat her visual field testing in six months.”

Case 4: 1 year later

- Patient presents with “cloudy vision”
- VA 20/24 OU
- Tdct OD 11.0 mmHg OS 12.2 mmHg
- Visual fields are as seen in the following slides- (bilateral altitudinal defects)
- Optic nerves are as seen in following slides (edema with hemorrhage OU)

Case 4

What is your tentative diagnosis
1. Cavernous sinus fistula
2. Temporal arteritis
3. Compressive neuropathy 2\textsuperscript{nd} to thyroid ophthalmopathy
4. Cavernous sinus stenosis

See discussion at end of handout
Case 1

• Diagnosis: fixed pupil secondary to repeated use of OTC drop containing anticholinergic agent

• Differential
  – Migraine, cluster headache, space occupying lesion, demyelinating disease, cavernous sinus syndromes, carotid dissection

Case 1- Pharmacologic Pupil

• General considerations
  – Patient age, health, presence or absence of accompanying signs, symptoms

• Differential- anisocoria
  – Pupils: evaluate in light vs. dark
  – Look for accompanying “P” signs: ptosis, proptosis, pain, poor movement/pareisis,
  – CT scan if collateral signs, symptoms
  – Pilocarpine test
Conducting Pilocarpine Test

• Pharmacologically dilated pupil
  • Larger than most other causes of anisocoria
    – 1% pilo will not constrict pupil
• Adies tonic pupil
  – Combine 1 drop 1% pilocarpine with 7 drops saline (1/8% pilo)
  – Instill 1 drop in affected eye and fellow
  – Hypersensitive pupil will constrict

Other Causes of Pharmacologic Pupil

• Pilocarpine
• Cocaine
• Tropicamide
• Scopolamine
• Alkaloids present in plants
  – Brugmansia, Delphinium, Jimson weed, deadly nightshade

Case 2

• This patient’s signs and symptoms resulted from:
  – Pituitary adenoma
  – Myasthenia gravis
    • 15% of patients with MG have associated thyroid disorders
Case 2

- Diplopia + ptosis + jaw claudication
- Pituitary mass (non-contributory)
- Diplopia in conjunction with other neuro signs and symptoms can signal serious conditions

**Diplopia Plus- Five to Fear**

- Pupil involvement suggesting third-nerve palsy
  - Cause: Aneurysm, usually of posterior communicating artery.
- Lid, pupil or eye movement alteration
  - Cause: Horner syndrome (ptosis with small pupil), third-nerve palsy (ptosis with big pupil), inflammation

**Diplopia Plus- Five to Fear**

- More than one cranial nerve palsy
  - Cause: intracranial or meningeal-based tumors, meningitis or polyneuropathy
    - Example CN III & VI due to cavernous sinus lesion
- Weakness or fatigue
  - Cause: myasthenia gravis- can mimic almost any ophthalmoplegia, cause aspiration or respiratory
Diplopia Plus- Five to Fear

- New kind of headache, scalp tenderness, pain with chewing
- Cause- giant cell arteritis
  - Can cause diplopia plus multiple other signs & symptoms

Myasthenia Gravis

- Nerve impulse triggers release of ACh acetylcholine into the synaptic cleft
- Binding of ACh to the postsynaptic receptors depolarises the muscle fibre
- Neuromuscular disorder (autoimmune) caused by antibodies to ACh receptors
  - Block action of ACh on postsynaptic receptors
- Key ocular symptoms
  - Diplopia
  - Ptosis

Myasthenia Gravis

- Ice test-apply ice pack to eyelid for @ two minutes. A positive response = “dramatic improvement” in the ptosis.
- Lab- elevated level ACh receptor antibodies- confirms the diagnosis
- Management-
  - Immunosuppresion- corticosteroids
  - AChE inhibitors- physostigmine, neostigmine
Case 3

- Fixed pupil secondary to herpetic-induced sphincter atrophy
- Rare but real


Case 3

- Objective: Report the association between (HSV) and iris atrophy with pupillary dilation
- Result: 13 of 13 patients demonstrated some degree of iris atrophy; 9 of 13 had pupillary dilation on the affected side despite not being on dilating drops.


Case 3

- Thirteen of 13 patients demonstrated some degree of iris atrophy; 9 of 13 had pupillary dilation on the affected side despite not being on dilating drops.
- “We suggest that unexplained pupillary dilation be added to the list of clinical clues suggesting a herpetic etiology of uveitis.”
Case 4

• Temporal arteritis resulting in:
  – Altitudinal defects
  – RAPD
  – Optic nerve edema with splinter hemorrhages

Giant Cell Arteritis

• First reported by Horton et al in 1937
• Vasculitis- affects medium & large caliber vessels
• Etiology-triggers unknown
  • Granulomatous inflammation of internal elastic lamina
  • Macrophages, lymphocytes, fibroblasts, and sometimes multinucleated giant cells
• Prevalence @15 per 100,000
• Sites: aorta, and branches, temporal and ophthalmic arteries, cranial nerves (optic)

Giant Cell Arteritis

• Most common symptoms of GCA- headaches, joint pain, facial pain, scalp tenderness, fever, anorexia - weight loss, and vision changes (reduced vision, diplopia)
• Most specific symptom- jaw claudication
  claudication = limping or lameness
• Affects females 2-4 times more frequently
• Age of onset after 50 years of age
• Average age of onset - 72 years of age
Giant Cell Arteritis: Lab findings

- Elevated erythrocyte sed rate (ESR)
  - Men: age/2
  - Women: age +10/2
- Elevated C-reactive protein (CRA)
- Asymmetric pulse or blood pressure
- Anemia
- Thrombocytosis—abnormally increased number of platelets in the

Giant Cell Arteritis

- Treatment:
  - Initially high dose steroids
  - Very gradual taper over months or years
  - Vision loss typically does not improve
  - Other signs, symptoms resolve over time