Common Neuro-ophthalmologic Conditions

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Disclosures

- no disclosures
Objectives

- Review presentation and differential diagnosis of more common neuro-ophthalmic conditions
  - Optic neuritis
  - Diplopia
  - Anisocoria
  - Idiopathic intracranial hypertension
31yo woman

- Right periobital pain/HA
- Right eye pain with eye movement
- Photophobia
- “no vision right eye”
- Decreased over 1-2 days

- Visual acuity
  - OD: Hand Motion
  - OS: 20/20

- Color vision
  - OD: unable
  - OS: 14/14 ishihara plates

- EOMI – painful OD
- Pupils: right RAPD
Funduscopic Exam

Right eye

Left eye
MRI brain wwo

T1 coronal wwo, with fat suppression
MRI brain wwo

T2 coronal, with fat suppression
Clinical presentation of Optic Neuritis

• **Pain**
  - 92% report peri/retro-orbital
  - Worse with eye movement
  - Typically presents before vision loss

• **Monocular vision loss**
  - 20/20 to NLP
  - 1/3 experience photopsias
  - Decreased color vision
    • Red desaturation
  - Visual field defects
    • Central
    • Peripheral

• **Pupils**
  • +Relative afferent pupillary defect

# Causes of Optic neuritis

## Most Common
- Idiopathic demyelination

## Other causes:
- Viral/bacterial
- Auto-immune
- Inflammatory
- Other
Optic Neuritis Treatment Trial (1992)

- Optic Neuritis Treatment Trial (1992)
- Compared the speed and extent of visual recovery

Patients treated with:
- Oral prednisone
  - increased risk of recurrent optic neuritis (35%)
- IV methylprednisolone
  - Risk of recurrent optic neuritis 16%
  - Decreased risk of developing clinically definite MS at 2 yrs (however, no difference at 3 yrs)
- Placebo
  - Risk of recurrent optic neuritis 17%

Beck et al 1992
### Outcomes

#### Optic Neuritis Treatment Trial (1992)

<table>
<thead>
<tr>
<th><strong>Visual</strong></th>
<th><strong>MS</strong></th>
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<tr>
<td>- Final outcomes at 12 months were the same with or without treatment.</td>
<td>- Predicted by baseline brain MRI.</td>
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<td>- Most begin to improve within 3-4 weeks.</td>
<td>- # of lesions at least 3 mm in size.</td>
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<td>- At 15 yrs</td>
<td>- If no MRI brain lesions – 25% risk.</td>
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<td>- 72% recovered 20/20 vision.</td>
<td>- If one or more white matter lesions - 72% risk.</td>
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<td>- 85% recovered 20/25 or better.</td>
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Atypical features of Optic neuritis

- Age greater than 50 yrs
- Optic disc pallor at presentation
- Bilateral simultaneous vision loss
- Absence of pain
- Pain and/or vision loss that progresses over weeks
- Poor visual recovery
- Associated systemic signs and symptoms
Sarcoid

- Pain is often absent
- Optic neuritis very steroid responsive

B) Irregular disc swelling-granulomas on the disc
Neuromyelitis Optica aka Devic’s disease

- NMO
  - Optic neuritis + spinal cord disease
  - Severe vision loss
  - CSF:
    - Neg OCB
    - Elevated Prot and WBC
  - Symptoms rarely improve
Para-infectious optic neuritis

Viral or bacterial

- 1-3 weeks after infection
- Children > Adults
- Often bilateral
- Prognosis: excellent
  - +/- steroids
- Cat scratch
- Syphilis
- Lyme disease
- HIV
- TB
- Post-vaccination

Macular star
Diplopia
## Diplopia

<table>
<thead>
<tr>
<th>Monocular</th>
<th>Binocular</th>
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<tbody>
<tr>
<td>• Ocular</td>
<td>• Cranial nerve paresis</td>
</tr>
<tr>
<td>• Dry eye syndrome</td>
<td>• Brainstem lesion</td>
</tr>
<tr>
<td>• Corneal</td>
<td>• Cerebeller lesion</td>
</tr>
<tr>
<td>• Refractive</td>
<td>• Intra-orbital mass</td>
</tr>
<tr>
<td>• Lenticular – cataract</td>
<td></td>
</tr>
<tr>
<td>• Retinal</td>
<td></td>
</tr>
<tr>
<td>• Psychiatric/functional</td>
<td></td>
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</tbody>
</table>
Cranial Nerve III Palsy

- Appearance of palsy
  - Extraocular movements
    - Eye goes down and out
  - Drooping eyelid
  - Dilated pupil
Cranial Nerve III Palsy

- CN III palsy has several potential etiologies
  - Intracranial process
    - Aneurysm!!
    - Tumor
    - Brain herniation etc
  - Ischemia

- Two Types
  - Pupil Involving
  - Pupil Sparing
Cranial Nerve IV Palsy

- Appearance of CN IV palsy
  - One eye is higher than the other
  - Patients will often compensate with a head tilt
- Patient will describe vertical double vision
  - May also note that 1 image is rotated
Cranial Nerve IV Palsy

- CN IV palsy potential etiologies
  - Trauma
  - Tumor

- Function of CN IV
  - Incyclotorsion
  - Infraducts eye
Cranial Nerve VI Palsy

- Function of CN VI
  - Abduction (moves eye out)
  - Eyes cross
  - Effected eye can not move out
Cranial Nerve VI Palsy

- CN VI palsy potential etiologies
  - Trauma
  - Ischemia
  - Tumor
  - Increased ICP
  - Cavernous sinus ICA aneurysm
Cranial Nerve Palsy

- **Testing/Treatment**
  - CT/CTA or MRI/A if suspicion for Intracranial process
    - Headache
    - Other neurologic symptoms
    - No microvascular disease
    - History of recent Trauma
  - If patient with microvascular disease (DM, HTN, Cholesterol, smoking)
    - No imaging required
    - Observation for 6-8 weeks
Anisocoria
Anisocoria greater in:

**Darkness**
- Physiologic (simple) anisocoria
- Horner’s syndrome

**Bright light**
- Adie’s (tonic) pupil
- Oculomotor (third) nerve palsy
- Pharmacologic mydriasis
Horner’s Syndrome

- Appearance of Horner’s
  - Small pupil
  - Slight drooping of eyelid
  - Anhydrosis
Horner’s Syndrome

• Sympathetic Pathway

• Etiology - variable
  • Lung tumor (Pancoast tumor)
  • Carotid dissection
  • Neck trauma
  • Cervical cord lesion
  • Lateral medullary syndrome (Wallenberg)
  • Viral/idiopathic
Adie’s (tonic) pupil

• Acute denervation of the postganglionic parasympathetic fibers

• Initially dilated

• Sluggish, segmental pupillary responses

• Better response to near effort
Causes of Adie’s pupil

- Idiopathic
- Viral ganglionitis
- Migrainous vasospasm
- Trauma
- Ocular surgery (e.g. scleral buckle)
- Tumors
Bilateral simultaneous tonic pupils

- systemic/autonomic peripheral neuropathy
- diabetes mellitus
- amyloidsosis
- syphilis
- paraneoplastic syndromes
- Sjogren’s syndrome
- polyarteritis nodosa
IDIOPATHIC INTRACRANIAL HYPERTENSION

- Idiopathic intracranial hypertension (IIH)
- Benign intracranial hypertension
- Pseudotumor cerebri
- Intracranial hypertension secondary to...
Clinical manifestations

- Headache
- Transient visual obscurations
- Pulse synchronous tinnitus
- Diplopia
- Visual loss

- Papilledema
- 6th nerve palsy
- Visual field defects
Papilledema

http://webeye.ophth.uiowa.edu/eyeforum/cases/case30/case30-Figure1.jpg
Papilledema

 Poorly Defined Borders

 Normal Disc
Papilledema with choroidal folds

“Champagne Cork”
Epidemiology

- Annual incidence
- General population 0.9/100,000
- Women 15 to 44 3.5/100,000
- Women 20-44 and 20% above ideal body weight 19.3/100,000
Epidemiology

- Before puberty boys = girls
- After puberty women affected 9 times as often as men
- Rarely develops in patients over 45
Almost all patients with IIH

- Daily, retro-bulbar, worse with eye movement
- Neck and back pain are prominent features
- Throbbing, nausea, vomiting, photophobia
- Often worse supine
Transient visual obscurations

- Brief episodes of monocular or binocular vision loss
- Partial or complete
- Likely due to disc edema leading to ischemia of the optic nerve head
Pulse Synchronous Tinnitus

- Pulsatile tinnitus 60%
- Unilateral or bilateral
- Abolished with LP or jugular venous compression
- Transmission of intensified vascular pulsations via CSF
**Diplopia**

- Unilateral or bilateral sixth nerve palsy
- Secondary to increased ICP
- Binocular horizontal diplopia
- Resolves when ICP lowered
Visual Loss

- Blurred vision
- Temporal dark spot
- Tunnel vision
- Profound or complete blindness
- Tempo variable: as soon as days
Other symptoms:

- Paresthesias
- Neck stiffness
- Arthralgia shoulders, wrists, knees
- Ataxia
- Facial palsy - rare
- Radicular pain
- Depression
Typical MRI findings of IIH

- Flattening of the posterior globe at the insertion of the optic nerve - 80% patients
- Empty sella - 70%
- Distension of the optic nerve sheath - 45%

Diagnostic criteria for idiopathic intracranial hypertension. Friedman and Jacobson. Neurology v 59 p. 1492-1495
Idiopathic intracranial hypertension

Lumbar Puncture

- Lateral decubitus position with legs relaxed
- 18- to 20- gauge spinal needle
- Document elevated CSF pressure
- Opening pressure > 250mm H\textsubscript{2}O
- 201 – 249 mm H\textsubscript{2}O are nondiagnostic
- Repeat LP may be necessary if initial OP nondiagnostic
- Rarely need 24 hour transducer monitoring through lumbar drain to diagnose
Workup for Contributing Factors/Mimics

- Vital signs - r/o hypertensive papillopathy
- Medications
  - Tetracycline
  - Retin-A, acutane
- Medical history
- Consider sleep study to eval for OSA
- Labs
  - Hct - anemia
  - TSH, free T4 - hypothyroidism
  - Vitamin D level
  - Parathyroid level - hypoparathyroidism
  - CMP
  - Specific tests for suspected conditions suggested by the history
Treatment

• Medical
  • Diet and weight loss
  • Medications
    • Diamox
    • Topamax

• Surgical
  • Optic nerve sheath decompression
  • CSF shunting