Best Neuro Lecture Ever

Cecelia Koetting, OD FAAO
Denver CO
Hines-Sight

Disclosures

Neurology as an Optometrist

• Signs and Symptoms noted on routine exams
• First place some patients go
• Aid in diagnosis and treatment
  ▪ Knowing when to order MRI, MRV, MRA
  ▪ Lumbar puncture/Spinal Tap
  ▪ Temporal artery biopsy
  ▪ Prescribing treatment
  ▪ Oral steroids
  ▪ IV steroids

Neuro can be confusing, so lets break it down into a few categories

• Cranial Nerve Palsies
• Optic Nerve Changes

Cranial Nerve Palsies

Cranial Nerve 3 Palsy

• Eye is turned “down and out”
  ▪ Abducens and superior oblique still function
• Eyelid is shut
  ▪ Levator palpebrae the lid retractor is paralyzed
• Possible pupil involvement
  ▪ Pupil sparing (normal reaction)
  ▪ Non-Pupil sparing (affected pupil is not reactive to light shone in either eye)

http://www.nature.com/eye/journal/v18/n3/fig_tab/6700625f1.html
CN 3 Palsy Causes

- Pupil involvement implies likely tumor/aneurysm (most concerning)
  - Something pressing on the nerve from outside
- Non-Pupil involvement implies likely diabetes/HTN/ischemia (most common)
  - Something affecting the nerve from inside
  ...sometimes

Whats the next step?

Diagnosis

- Testing
  - In office
    - Versions/Ductions/Prisms
    - EOM, Ductions and versions
    - Forced Ductions
  - Outside office
    - No pupil involvement
      - BS/blood work for DM
      - BP
      - Monitor for future pupil involvement
    - Pupil involvement
      - Immediate MRI of head and orbit and MRV with and without contrast

Versions and Ductions

How to Document

Forced Ductions

Prism Measurement of Deviations

Cranial Nerve 4 Palsy
CN 4 Palsy
- CN 4 Controls superior oblique extraocular muscle
- Upward deviation of affected eye
- Cyclotorsion of the eye
- Head tilt away from lesion
- Diplopia

CN Palsy 4 Causes
- Trochlear nerve is the longest and skinniest, susceptible to injury
- Most common causes
  - Trauma
  - Old age
  - Congenital
  - Ischemic
  - Tumor

Diagnosis
- A good, detailed history
- Blood pressure, blood sugar
- Possible blood work (cholesterol, A1C)
- Parks 3 step
- MRI of head and orbit with and without contrast

Parks 3 Step
- Using cover test to localize the muscle affected
  1. Which eye is higher in primary gaze?
  2. Is it worse in right or left gaze?
  3. Is the hypertropia worse with right or left head tilt?

Treatment
- Treatment based on findings:
  - Congenital and trauma related: follow up with neurologist
  - Lesion/mass refer quickly to neuro surgeon
  - High BP/VP monitor and contact primary care doctor to help
- Continue to monitor the patient

Case #1
- 63 year old AA woman with chief complaint of new onset diplopia stable over the last 2 months
  - Images are side-by-side and occasionally triple
  - Resolves when right eye is covered
  - Increased headaches
- Neurologist seen 2 months prior with normal MRI per patient
How we feel inside...

**History**
- No prior ocular history of surgeries or strabismus
- Medical history includes lung cancer in 2005 with relapse in 2014
- Former smoker, quit 12 years prior
- Hypertension and hyperlipidemia controlled with medication
- NOT diabetic or borderline DM

**Exam Findings**
- BCVA 20/20 OD and OS
- Red Cap and color vision normal
- No APD
- Version andduction test reveals a complete loss of abduction OD and full range of motion OS
- SLE and DFE show no other abnormalities

---

**What do we do next?**

**In Office**
- Forcedduction test OD
  - Negative
- Blood pressure
  - 140/80
- Dx: Cranial Nerve VI palsy

**Further Testing**
- Blood work including CBC, A1C, ESR, and CRP
  - All negative
- Same day MRI and MRV of head and orbit with and without contrast

- Compared with her MRI two months prior, a sizable (2.1x1.6x2.4cm) peripheric lesion abutting the right cavernous sinus and involving right Meckel’s cave was detected
- Two smaller enhancing brain nodules were found in the left parafalcine occipital lobe and in the left frontal lobe
- All were considered suspicious for lung cancer metastases.
How to order an MRI

• If it is emergent (in the case of possible Optic Neuritis or AION):
  o Refer to local ED within 24-48 hours for MRI
  o Can send with a written script for MRI of head and orbits with and without contrast
  o Include why you are ordering it
  o Sudden decrease in vision OD with pain, possible optic neuritis
  o Include a phone number to reach the doctor and be ready for a call
    • They will likely ask for treatment suggestion if confirmed diagnosis
  o Can send with standing order for how to treat if positive diagnosis

• Sudden decrease in vision OD with pain, possible optic neuritis
  o Include a phone number to reach the doctor and be ready for a call
    • They will likely ask for treatment suggestion if confirmed diagnosis
  o Can send with standing order for how to treat if positive diagnosis

How to order an MRI

• In a non-emergent situation (papilledema likely IIH):
  o Order an MRI of the head and orbits with and without contrast within a few weeks
  o Can be scheduled with out patient clinics or at MRI centers
  o Your front desk staff can help the patient with this.
    • MRA vs MRV
      o Artery vs veins
      o Aneurysms, dissections, cerebral venous sinus thrombosis

When is CT helpful?

• Good to view:
  o Fractured skull
  o Cardiac causes
  o Early involvement from soft tissue mass
  o Metallic foreign body
  o Post-infection

• Indicated when:
  o Orbital fracture
  o Proptosis, swelling of orbital tissues (cellulitis, abscess, etc)
  o Spontaneous subdural hematoma
  o Intracranial or intramedullary foreign bodies
  o Cranial palsy (post stroke MRI)

• Avoid if possible in pregnant patients

Treatment

• Spoke with Neurologist that day, referred to neurosurgeon
• Patient underwent five rounds of radiation and came in every four weeks for versions/duction testing and visual fields

CN 6 Palsy

• CN 6 controls the lateral rectus muscle
  o Loss of abduction
  o Also have head 180
  o Double vision

CN 6 Palsy Causes

• Congenital
• Increased intracranial pressure
• Idiopathic intracranial hypertension
• Meningitis
• Head injury
• Tumor
• Ischemic (HTN, DM)
Diagnosis

- EOM, Ductions and versions
  - Forced duction
- Good history, BP and BS
  - Possible blood work, A1C cholesterol
- MRI of head and orbit
  - With and without contrast

Treatment

- Treatment based on findings
  - Lesion/mass refer immediately to neuro surgeon
  - High BS/BP monitor closely
  - If no improvement within 1 month obtain MRI if have not
  - Work closely with PCP to treat underlying cause

Optic Nerve Changes

Optic Nerve Head Edema

Unilateral ONH Edema

- Retinal vein occlusion
- Optic neuritis
- Articic ischemic optic neuropathy
- Non- Articic ischemic optic neuropathy
- Diabetic papillitis

Case #3

- 30 year old white female
- ICC: unilateral sudden decrease in vision in her right eye noted yesterday morning. No improvement today. States she feels some pain when she moves her eyes.
- DVA OD 20/100, PH N; OS 20/20
- EOM: OD FROM (+) pain on movement in all gazes; OS FROM (-) pain/diplopia

- Red cap test: OD 20%; OS 100%
- Color Vision Ishihara: OD 5/17 plates; OS 17/17 plates
Now what?

Optic Neuritis
- Inflammation of the optic nerve that damages the optic nerve tissue
- 6.4 per 100,000 in US
- Unilateral in 70% of cases
- 3:2 female:male
- Most often 30's, range 20-60 YOA
- Triad
  - 1. loss of vision
  - 2. dyschromatopsia
  - 3. eye pain (worse with movement)
- Optic disc swelling
- (+) APD
- Orbital MRI will show inflammation of ON

Optic Neuritis
- Visual field loss will usually occur
- Red cup and color vision changes
- Red cup destruction and color vision deficiencies in the affected eye
- Pupil APD in affected eye may occur

Optic Neuritis
- Treatment
  - IV steroids followed by oral steroids
  - After treatment initiated, 30-2, color vision, and OCT-G should be performed to monitor improvement
  - Patient should be referred to a Neurologist for risk of ON association with MS

Optic Nerve Treatment Trial
- 448 Patients with optic neuritis seen within 8 days of symptom onset
- Improved visual prognosis with IV steroids vs oral vs placebo
- Primary outcome showed those treated with IV methylprednisolone followed by oral steroids led to faster visual recovery
- Patients treated with vision 20/30 or worse at presentation had the best recovery
- Decreased rate of re-occurrence with use of oral steroids alone
- 14% IV vs 20% oral or placebo

Multiple sclerosis
- Immune-mediated process directed against the CNS
  - Attacks the myelin and the nerve fibers
  - Visual field defects
  - Bilateral internuclear ophthalmoplegia (INO)
  - Brainstem and cerebellum lesions
  - Optic Neuritis
    - 75% occurrence, initial symptom in 14-20%
MS Risk After Optic Neuritis

- High risk of developing MS (50%)
  - 15-year risk of MS was 50% overall
  - 25% risk of MS when MRI is normal
  - 75% risk of MS when MRI shows one or more lesions

Neurolyelitis Optica (NMO)

- Previously thought of as variant of MS
- Demyelination of optic nerve and spinal cord
- Associated with aquaporin-4 (a water channel present in glial cells) antibodies.
- Testing for NMO-IgG should be considered in those patients with bilateral ON or ON coupled with longitudinally extensive transverse myelitis (LETM), recurrent ON, or brain MRIs atypical for MS
- No cure, but similar treatment to MS
- Poor prognosis, loss of muscle function, often death occurs 2/3 respiratory complications

Bilateral ONH Edema

- Papilledema
- Idiopathic intracranial hypertension
- Optic nerve pseudoequema

Case #4

- 25 year old African American female
- CC: Increase in headache frequency and intensity over the last few months
- (+) weight gain over the last 6 months
- (-) tinnitus or birth control use
- DVA OD 20/20; OS 20/20

Now what?

MRI

- MRI of the head and orbit with and without contrast
  - MRI of head important for masses, lesions and will check the posterior portion of the brain for Chiari malformation
  - May come back normal with no findings
- Indications of elevated CSF pressure
  - Posterior globe flattening
  - Empty sella
  - Increased tortuosity of optic nerve
  - Enlarged optic nerve sheath
Neuro consult
- With a normal MRI or with findings that indicate likely IIH the next step is a neurology consult
- They will usually order a lumbar puncture to confirm the exiting pressure of CSF and if any infection present
- May order an MRV

Papilledema
- Bilateral swollen optic nerves secondary to increased intracranial pressure
- OCT-G and 30-2 HVF
- Most common VF defect
  - Enlarged blind spot
  - Peripapillary scotoma
  - Often no visual field defect
- Quickly accompanied by and MRI of head and orbit to rule out space occupying lesion
- Must be confirmed with a lumbar puncture to check the ICP

Idiopathic Intracranial Hypertension
- AKA Benign Intracranial Hypertension and pseudotumor cerebri
- Increased intracranial pressure with unknown cause
- Diagnosis
  - Signs and symptoms
    - Headaches, tinnitus, tingling in fingers and toes
  - Diagnostic:
    - Within 1-2 weeks
    - Lumbar puncture
    - Increased exiting pressure with normal fluid
    - Repeat puncture
- Usually not treated

IIH
- Causes
  - Weight
  - Birth control
  - PCOS
  - Minocycline, doxycycline, etc
- Long term concerns and treatment
  - Glaucoma/ONH damage
    - Monitor with OCT-G
  - Diamox (acetazolamide)
  - Topamax
  - Shunt
  - Optic nerve fenestration
  - Weight loss
- Approximately 10% body weight loss has been shown to reverse

Co-managing
- Monitor the patient closely along with neurology
- Patient sees neurology within a month for remaining testing, diagnosis, and treatment
- Should see the patient back within 1-2 months of neurology for repeat OCT-G and 30-2 to monitor
- Follow patient every 3-6 months for repeat testing to aid neurologist in determining if medication is working adequately.

Pseudoedema
- Hyaline bodies that become calcified and are located within the optic nerve head.
- Approximately 3-4/1000 people
- As drusen become larger over time they can cause a progressive visual field defect due to the secondary thinning of the RNFL.
- Confirmation gold standard is obtained with B-scan or CT
  - Can also perform a fluorescein angiography to confirm
  - Monitor with OCT over time
ONH Drusen
- 24.87% of ONHD have a visual field defect
- Most common visual field defect
  - Inferior nasal step
  - Sectoral arcuate scotoma
  - Enlarged blind spot
  - Concentric peripheral constriction

Treatment
- Monitor with visual fields and OCTG
- If vision becomes compromised can treat with topical IOP lowering medications
  - Secondary glaucoma

Case #5
- 65 year old white male
- CC: Noted a sudden decrease in vision in his left eye this morning when he woke up. Pt denies any pain, but he noted a slight throbbing in his temple.
- BC DVA OD 20/20; OS CF @5
- 3+ APD OS
- DFE: OD ONH WNL; OS ONH 3+ edema, hyperemia

MRI and Blood Work
- MRI of head and orbit with and without contrast within 24 hours
  - Normal no findings
- Blood work
  - CRP and ESR
    - Both elevated

Now What?
Temporal Artery Biopsy

Arteritic Ischemic Optic Neuropathy
- Arteritic [Giant Cell-Arteritis/ Temporal arteritis]
  - Vasculitis within the medium and small sized arteries around the head, primarily over 60 YOA
  - No reports of GCA in any patient under the age of 50 YOA
- Signs and symptoms
  - Sudden painless vision loss
  - Scalp tenderness/headache
  - Jaw claudication, especially while eating
  - Polyarthalgia of arm and shoulders
  - Fever, night sweats, weight loss

Blood work
- Elevated Sedimentation Rate (ESR)
  - Elevates in response to acute and chronic inflammation
- C-Reactive Protein (CRP)
  - An acute phase protein that increases quickly with inflammation and decreases faster than ESR with resolution
  - Do not start oral steroids until after blood work

Temporal Artery Biopsy
- Used to help confirm diagnosis of AION
- Skip lesions can occur decreasing the test's accuracy and specificity
- Sometimes initiate treatment even without a positive TAB
- Do not start oral steroids until after blood work
- May initiate treatment of oral steroids up to 72 hours prior to biopsy if highly suspect after blood work results
- These need to be managed by the patient's PCP
- Long term treatment of up to a year with oral steroids

Non-arteritic Ischemic Optic Neuropathy
- Localized ischemic event at junction of optic nerve
- May be younger in age than AION (40-60 YOA)
- Signs and symptoms
  - Sudden painless vision loss
  - VA decreased
  - Less severe than AION
- Signs
  - APD
  - Pale disc swelling
  - Flame shaped heme

NAION
- Diagnosis of exclusion
  - Normal MRI
  - May find chronic microvascular changes on MRA
  - Normal ESR/CRP
  - 40% show some improvement in vision over the next 6 months
  - Monitor with visual fields
  - Optic nerve edema will resolve within 8 weeks
  - Can monitor with OCTG
  - Risk of contralateral eye involvement

NAION Treatment
- It has been suggested in a study by Foulds in the 1970's that the patients may benefit from long-term visual recovery from the use of 40-60mg of oral prednisone for 1 month.
  - 85% of patients treated with 60mg oral prednisone showed visual acuity improvement compared to those untreated
- NAION Treatment
- More recent study, 2008, Hayreh and Zimmerman 696 eyes
  - Treated within 2 weeks of onset with 70mg oral prednisone tapered
  - 69.8% of eyes treated had an improvement in visual acuity
  - Only 40.5% of eyes untreated had an improvement in visual acuity
Levodopa

Levodopa for NAION

- 59 patients within 15 days of onset NAION
  - Either untreated or given 25mg carbidopa/100mg levodopa PO TID
  - 19/23 in the levodopa group BCVA improved and none got worse
  - 6/14 in control group BCVA improved and 4/14 got worse

Summary

- It is important to find the underlying cause of double vision and/or swollen optic nerve and to start testing early
- Optometrists can work alongside neurology to manage these patients

Questions?

Thank you!