Neuro-Ophthalmology and the Healthcare Team

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1. Vision loss - acute and chronic
   - Corneal defects
   - Retinal changes
   - Lens displacement or related changes

The EYE Examination

- Check vision
  - Eye chart
  - Distance vision
  - Near vision
  - Presbyopia – check age + lens

Vision

- A subjective disorder with objective findings.
  - The presence of the rAPD
  - Changes in the back of the eye
    - Pallor
    - Edema
    - Inflammation
Tools for vision testing

- Direct light
- Vision cover/uncover
- Eye/vision chart
- Color vision
- Visual field testing: confrontational, automated
- MRI/MRA/CT
Retinal Physiology

- Cones
- Rods

Swinging light test

- First have the patient look into the distance
- Next determine the reactivity of the pupils
- Next shine the light into the eyes; right no more than 3 seconds then the contra lateral eye.

Normal is no change in the contra lateral pupil initially when contacted by the light.

Abnormal is the dilation of the contra lateral pupil when contacted by the light.

Fundus Examination

- Direct
  - Standard
  - Pan optic
    - Both to review the vascular changes
    - Both to review the optic nerve
- Slit Lamp
  - Ophthalmologist/optometrist
**Visual fields**

- Confrontational
  - Operator/examiner
- Automated
  - Humphrey
  - Goldmann

**Visual Field/Ophthalmic Terms**

- Arcuate
- Monocular
- Binocular
- Chiasmal
- Postchiasmal
- Bitemporal hemianopia
- Congruous
- Incongruous

**Field/Ophthalmic Terminology**

- Scotoma: depression of sensitivity to surroundings
  - Central
  - Paracentral
  - Cecocentral
Patient #1

- 56 year old
- Presents with sudden vision loss in the left eye
  - There is pain
  - No pain on eye movement
- Examination
  - Noted rAPD
  - Pattern of loss on confrontation
  - FUNDUS: swollen nerve optic

#1

- Labs
  - Sedimentation rate normal
  - ?
  - ?
  - MRI ?
  - Vitals
    - Blood pressure ?

Anterior Ischemic Optic Neuropathy

- Loss of peripheral and/or central vision
- Optic disc swelling
- Peripapillary swelling
- Common disorder
- Usually are 50-60
- Inferior hemisphere most common affected

NAION

Pallor
Cont.........

- Risk factors: small cup-to-disc
- Diabetes or hypertension

Patient #2

- 72 year old
- Presents with sudden vision loss in the left eye
  - There is pain
  - No pain on eye movement
- Examination
  - Noted rAPD
  - Pattern of loss on confrontation
  - FUNDUS: swollen nerve-optic

#2

- Labs
  - Sedimentation rate elevated
  - ?
  - ?
  - MRI ?
  - Vitals
    - Blood pressure ?

Evaluation continued:

- Consider Bx?
  - ? Bilateral or single
    - If negative
      - Then what—

Arteritic AION

- Age onset 70-80’s
- Vision loss seems more severe as a general rule
- Jaw claudication, temporal headache, weight loss, fatigue and joint pain.
- Lupus, polyarteritis nodosa, syphilis, and radiation necrosis

Cont......

- ESR
- C-reactive protein
- TA bx- suggest at least a one inch sample
Patient #3

- 36 year old
- Presents with sudden vision loss - OU
  - There is pain
  - No pain on eye movement
Examination
  Noted no - rAPD
  Pattern of loss on confrontation
  FUNDUS: swollen nerve; optic

IIH/PTCs

- Child bearing age
- Bilateral
- Nerve – swelling, maybe edema (papilledema)
- Family Hx
- Medication effect
  - treatments

#3

- Labs
  - Sedimentation rate normal
  - ?
  - ?
  - MRI ?
- Vitals
  - Blood pressure ?
Patient #3

- Fundus:
  - Nerves abnormal
  - Small-cup-to-disc ratio
  - Asymmetric
  - Why elevated pressure
  - New meds, trauma, non-compliance etc.

Patient #4

- 28 year old
- Presents with sudden vision loss
  - There is pain
  - Pain on eye movement
- Examination
- Noted aRPO
- Pattern of loss on confrontation
- FUNDUS: no swollen nerve optic

Optic Neuritis

- Vision loss: acute and chronic
- Acute vision loss
  - Optic neuritis
  - Ischemic/compressive/inflammatory
In this slide, we discuss the diagnosis and signs of Multiple Sclerosis (MS).

**Diagnosis**
- History
- Neurologic
- MRI
- CSF

**Signs of Multiple Sclerosis**
- Spasticity
- Hyperreflexia
- Babinski's
- Dysmetria or tremor
- Nystagmus and diplopia
- Impaired vibratory
- Facial weakness
- Impaired pain/touch
- Impaired temperature
- Changes in intellectual function

Diplopia is also mentioned, but the context is not clear from the image.
Definitions:
- Diplopia
  - Double vision
  - Monocular or Binocular?
  - Horizontal or vertical or oblique
  - Constant or intermittent
- Presentation
  - Sudden or chronic
  - Associated events or conditions

Terms – related- maybe
- Blurred vision
  - Near near
  - Things look off
  - I can not read
  - I can not drive

Visual
- The examination
  - Ductions
  - Versions

Monocular Diplopia
- Keratoconus
- Lens opacities
- Retinal pathology
  - Macular pathology

Divergence Insufficiency
- Deviation
  - Old 6th
  - Bilateral 6th
**Incomitant Deviations**

- Fusional amplitudes
  - About 2 diopters vertical
  - About 10-20 diopters horizontal

**Retrictive Misalignment**

- Thyroid orbitopathy
- Post traumatic
- Orbital myositis
- Neoplasm
- Congenital

**Thyroid - Orbitopaty**

- Most common
- Inferior and medial rectus most common
- Proptosis, chemosis, lid retraction and lid lag

- Evaluation - *forced ductions*. MRI- should reflect muscle change, but **no** tendon change

**Trauma**

- If at the most posterior aspects of the orbital region—near the cone—narrow region
- Blowout fractures
  - Inferior orbital floor-entrapment—inferior rectus
  - Medial rectus—less likely

**Orbital myositis**

- Ophthalmoplegia and pain
  - Conjunctival hyperemia, chemosis, maybe proptosis

- Usually found on MRI
  - NOTED tendon and muscle enlargement
  - Maybe related to lupus, sarcoidosis or Wegener’s granulomatosis

**Neoplasms**

- Meningiomas
- Mets
Congenital

- Brown syndrome
  - Limited upgaze, when eye is adducted
  - Short superior oblique tendon

- Fibrosis syndrome
  - Agenesis – ocular motoneurons in brain stem (superior division, 3rd cranial nerve – atrophy superior rectus and levator palpebral muscles)
  - CFEOM – congenital fibrosis of the extraocular muscles
  - Autosomal dominant – bilateral ptosis and external ophthalmoplegia

- Moebius syndrome
  - Agensis – 6th nerve nuclei and 7th nerve nuclei
  - Facial diplegia and ophthalmoplegia.
  - May also have scoliosis and atrophy of the tongue

- Duane Retraction Syndrome
  - Failure formation of the 6th nerve nucleus
  - Abducens paresis
  - Aberrant co-contraction of the horizontal rectus muscle
  - Anomalous branch of the 3rd nerve w/ the orbit
  - So, attempted adduction of the eye will trigger retraction of the eye, narrowing of the lid fissures.
  - May be bilateral. Do not report diplopia

Internuclear

- INO
  - Medial longitudinal Fasciculus (MLF) connecting the 6th nerve nucleus on one side medial rectus subnucleus (3rd) on the contralateral side
    - Slow adducting saccadic velocity nystagmus of the abducting eye. May have limited movement of the adducting eye. May have a gaze diplopia – paralytic strabismus

- One-and-a-half syndrome
  - Pontine lesion big enough to affect the MLF and the PPRF or 6th nerve nucleus on the same side
    - Move only abduction of the contralateral eye (stroke)

- Bilateral INO
  - MLF dual lesion, but also have gaze evoked nystagmus in upgaze –vertical vestibular nuclei (stroke)

3rd nerve

- Nuclear
- Fascicular
- Subarachnoid
- Divisional
- Aberrant

Nuclear 3rd

- Damage to one 3rd nerve nucleus
  - Bilateral ptosis, bilateral dysfunction of both superior rectus muscles and the usual pupillary issues and eye movement abnormalities.

- One and a half syndrome
  - Notch on cerebellum just after they leave subcortical therefore exist in the mid line (stroke)

Fascicular 3rd

- Cranial nerve injury beyond nucleus, but still in brainstem
  - Weber syndrome
    - Damage superior cerebellar peduncle – contralateral hemisphere

- Benedikt syndrome
  - Damage tectum, substantia nigra – contralateral midbrain

- Claude syndrome
  - Damage dorsal midbrain, superior cerebellar peduncle – contralateral area

- Nothnagel syndrome
  - Damage dorsal, motor area plus 2nd cranial – superior colliculus eye movements and arrest
Subarachnoid
- Brainstem to the cavernous sinus
  - Cause is usually microvascular injury to the nerve
  - Risk factors
    - DM, HTN, lipids

3rd nerve palsy - complete
- Ptosis, downward, outward deficit and pupil dilated
  - microvascular injuries, compression, infiltrative disorders, increased intracranial pressure

Pupil- involving 3rd nerve palsy
- If incomplete
  - concern for compression
    - Aneurysms
      - Post. Communicating/internal carotid

Pupil- sparing 3rd nerve palsy
- Complete loss of lid function and movement from the 3rd nerve
  - microvascular
    - Usually DM

Pupillary Dysfunction
- Normal lid and extraocular movements
  - Usually benign
    - Adie tonic
    - Pharmacologic

Divisional 3rd
- Two divisions in the cavernous sinus
  - Superior
  - Inferior

Usually other cranial nerves are involved – rare in isolation
4th nerve

- Worse in down gaze and head tilt

Transient vision loss

The approach to this clinical situation:

- What are the question:
  - Is the vision loss monocular or binocular
  - How old is the patient
  - What was the duration of the visual loss
  - What was the pattern of visual loss and recovery

- Vascular
  - Carotid artery or retinal circulation or optic nerve
  - Posterior circulation or migraine - visual cortex
  - Younger than 50 - migraine, vasospasm
  - Pregnant - eclampsia
  - Older than 50 - consider TIA, GCA

- Monocular loss
  - Duration 1-10 minutes 70-90% ipsilateral carotid
  - Vision loss lasing seconds - optic disc drusen, papilledema

- Shade coming down
  - TIA
  - Uhthoff syndrome
  - Vasospasm/migraine
  - Fortification spectra's
  - Optic neuroticism
Transient Monocular vision Loss

- Determine the status of the afferent visual axis
- Best corrected vision
- Visual fields
- Afferent pupillary defects
- Fundus exam
- What to look at: atrophy, vascular abnormalities, or orbital disease

Transient Monocular vision Loss

- Blepharospasm
- Cannot keep the eyes open
- Irregularity of the corneal tear film
- Testing: slit lamp reveals abnormal appearing film and cornea. Rapid tear film break down and punctal keratopathy
- Schirmer's

Transient Monocular vision Loss

- Ocular continued
- Mydriasis
- UCH
- Large PVD
- Angle closure glaucoma
- Macular degeneration - delayed vision recovery - exposed to light
- Gonioschisis
- Retinitis pigmentosa
- Choroidal - chronic high myopia and colobomas

Transient Monocular vision Loss

- Orbital
- Hemangioma
- Meningioma
- Intraconal mass
- Positional maybe with looking down

Transient Monocular vision Loss

- Systemic
- Retinovascular
- Cardiovascular
- Amaurosis fugax
- Sudden painless
- Lasting 2-30 minutes

Transient Monocular vision Loss

- Embolic
- Retinal circulation
- Painless vision loss
- Lasting 2-30 minutes
- Three types
- Cholesterol
- Platelet-fibrin
- Calcium
**Transient Monocular vision Loss**

- Other cause - Hypercoable states, mitral valve prolapse, cardiac arrhythmias, GCA, vasospasms-migraine, HTN, DM, CADASIL.

**Transient Monocular vision Loss**

- Laboratory evaluation:
  - Angiogram
  - >1% with experience
  - MRA
  - Carotid dissection
  - Echocardiogram
  - -valves

**Transient Monocular vision Loss**

- Risk for vision loss

**Transient Monocular vision Loss**

- Vasculitis
  - Usually over 50 years old
  - -sed rate C-reactive protein
  - Hypoperfusion
  - Ocular ischemic syndrome

**Transient Binocular vision Loss**

- Migraine: Most common cause

- Occipital mass/lesion: Fixed deficit – unilateral or central

**Transient Binocular vision Loss**

- Ocular ischemic syndrome