Neuro-Ophthalmology

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Lecture Content

- Disorders of optic nerve and retina
- Chiasmal and retrochiasmal disorders
- Pupil disorders
- Motility disorders
A 65-year-old woman has abrupt onset of visual loss in the right eye and weakness in the left arm and leg. There is some tingling of the left hand and left corner of her mouth. She is most likely to have a plaque in which blood vessel:

- A. Anterior cerebral artery
- B. Basilar artery
- C. Internal carotid artery
- D. Middle cerebral artery
- E. Posterior cerebral artery
A 35 year old woman woke with double vision. She reports that it gets better if she covers one eye. On exam: when looking to the left, her left eye has nystagmus to the left and her right eye does not move past the midline. She is able to look right without difficulty. She has normal vertical eye movements and is able to converge. The rest of her neurologic exam is normal. The most likely diagnosis is:

- A. Cavernous sinus thrombosis
- B. Midbrain lacune
- C. Multiple sclerosis
- D. Myasthenia gravis
- E. Thyroid ophthalmopathy
Question #3

- A 23 year old woman has the following visual field deficits.
- Where is the lesion?

- A. Optic radiation in the temporal lobe (Meyer's loop)
- B. Optic nerve at the chiasm
- C. Optic radiation in the parietal lobe
- D. Lateral geniculate nucleus
- E. Calcarine cortex of the occipital lobe
Question #4

- A 60 year old man is unable to report seeing a comb and a fork at the same time even when the 2 objects are held next to each other. He also has poor coordination of hand and eye movements, and difficult tracking objects visually. What is the most likely diagnosis:
  - A. Holmes-Adie syndrome
  - B. Balint syndrome
  - C. Gerstmann syndrome
  - D. Parinaud’s syndrome
  - E. Malingering
A 54 year old man has double vision. On exam, the left eye is deviated down and out. He also has ptosis and mydriasis. What is the most likely diagnosis:

A. Myasthenia gravis
B. Guillain-Barré syndrome
C. Posterior communicating artery aneurysm
D. Kearns-Sayre syndrome
E. Horner syndrome
Disorders of Optic Nerve and Retina

- Drusen
- Optic neuritis
- Papilledema
- Retinal emboli
- Retinal infarcts
Optic Nerve Drusen

- Refractile bodies
- Frequently present at the nasal margin. Visual field defects are common in patients
- Afferent pupillary defect in asymmetric cases
Pseudotumor Cerebri

- **Symptoms**
  - Headache
  - Papilledema
  - Stabismus

- **Evaluate**
  - Brain MRI with MRA and MRV
  - Lumbar puncture with opening pressure

- **Treatment**
Membrana limitans interna
Stratum opticum
Ganglionic layer
Inner plexiform layer
Inner nuclear layer
Outer plexiform layer
Outer nuclear layer
Membrana limitans externa
Layer of rods and cones
Pigmented layer

Fibers of Müller
neuronal layers of retina

pigment epithelium
photoreceptors
horizontal cells
outer synaptic layer
bipolar cells
amacrine cells
inner synaptic layer
ganglion cells
optic nerve fibre
optic nerve

http://www.insightseyecare.net/EyeEducation/retinaNeuronalLayers.jpg
This refractile, yellow fleck is a platelet-fibrin-cholesterol retinal embolus called a Hollenhorst plaque.
Chiasmal and Retrochiasmal Disorders
## Autonomic Control of Pupils

<table>
<thead>
<tr>
<th>Target</th>
<th>Sympathetic</th>
<th>Parasympathetic</th>
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<tbody>
<tr>
<td>Pupil dilator muscle</td>
<td>Alpha 1 contracts</td>
<td>M 3 relaxes</td>
</tr>
<tr>
<td></td>
<td>• Mydriasis</td>
<td>• Miosis</td>
</tr>
<tr>
<td>Ciliary muscle</td>
<td>Beta 2 relaxes</td>
<td>M 3 contracts</td>
</tr>
<tr>
<td></td>
<td>• Long range focus</td>
<td>• Short range focus</td>
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Pupil Disorders

- Afferent pupillary defect (APD)
- Horner
- Tonic
- Third nerve palsy
Marcus-Gunn Pupil or Afferent Pupillary Defect

- Optic neuritis
- Ischemic optic neuropathies
- Glaucoma
- Traumatic optic neuropathy
- Radiation optic nerve damage
- Miscellaneous optic neuropathies, such as Leber's optic neuropathy
- Ischemic retinal disease
Marcus-Gunn Pupil or Afferent Pupillary Defect

- Retinal detachment—if the macula or at least two quadrants of retina
- Severe macular degeneration
- Not with cataract
Argyll Robertson Pupil

- Constrict when the patient focuses on a near object (they “accommodate”)
- Do NOT constrict when exposed to bright light
- Neurosyphilis
Horner’s Syndrome

- Ptosis (superior tarsal muscle or Müller's muscle)
- Miosis
- Anhydrosis
- Dilation lag
- Enophthalmos
- Iris hypopigmentation in congenital Horner’s due to interference with melanin pigmentation
- Lesion of cervical or thoracic sympathetic chain
- Pancoast tumor (apical lung tumor)
Testing in Horner’s Syndrome

- Cocaine test
  - Blocks the reuptake of norepinephrine
  - Normal pupil dilates
  - Horner’s: no dilation
  - Apraclonidine (alpha antagonist) reversal of miosis on affected side

- Paredrine test
  - Hydroxyamphetamine causes neurotransmitter vesicle release if 3rd order neuron is intact which results in mydriasis
Holmes-Adie Syndrome or Ross’s Syndrome

- Mydriasis
- Loss of deep tendon reflexes
- Diaphoresis
- Accommodative paresis
- Photophobia
- Ciliary and spinal ganglia
- 1/8% pilocarpine test
Third Nerve Palsy

- Down and out (infraducted and abducted)
- Ptosis (levator palpebrae some bilateral innervation)
- Mydriasis (iris sphincter from ipsilateral Edinger-Westphal subnucleus)
- Posterior communicating artery aneurysm
Motility Disorders

- Abnormal movement
  - Nystagmus
  - Other
- Cranial nerve III, IV, VI paresis
- Parinaud’s
- Supranuclear gaze palsy
- Internuclear ophthalmoplegia
- One and a half syndrome
Nystagmus

- Periodic rhythmic ocular oscillation of the eyes
  - Equal amplitude and velocity (pendular nystagmus)
  - Slow initiating phase and a fast corrective phase (jerk nystagmus)

- Horizontal nystagmus
- Vertical nystagmus
- Rotary
- Congenital nystagmus
- Acquired nystagmus
Other Abnormal Eye Movements

- Ocular bobbing
- Ocular flutter
- Opsoclonus
Lateral view

- Trochlea
- Lateral rectus
- Superior rectus
- Superior oblique
- Optic nerve
- Inferior rectus
- Inferior oblique
- Lateral rectus

Superior view

- Medial rectus
- Superior oblique
- Superior rectus
- Optic nerve
- Inferior rectus
- Inferior oblique
- Lateral rectus
- Annulus of Zinn
- Medial rectus

- Trochlea
- Superior oblique
- Superior rectus
- Lateral rectus
- Medial rectus
- Inferior oblique
- Inferior rectus
Paresis of Individual Ocular Muscles

- Graves disease
- Orbit pathology
- Cavernous sinus pathology
- Myasthenia gravis
- Guillain-Barré syndrome
- Mitochondrial disease
- Myopathies
Fig. 18.73
Parinaud dorsal midbrain syndrome. (a) Defective upgaze; (b) straight eyes in primary position; (c) normal downgaze
(Courtesy of D. Thomas)

Fig. 18.74
T1-weighted MRI scan showing a pinealoma. (a) axial view; (b) sagittal view—note dilated ventricles
(Courtesy of D. Thomas)
Parinaud’s or Dorsal Midbrain Syndrome

- Paralysis of voluntary upgaze
- Accommodative paresis
- Convergence—retraction nystagmus
- Eyelid retraction (Collier’s sign)
- Conjugate down gaze in the primary position
- Pineal mass
Progressive Supranuclear Palsy

- Gait and balance problems
- Difficulty with downgaze
- Mood and behavior problems
  - Depression and apathy
- Dementia
Internuclear Ophthalmoplegia

- Lack of conjugate adduction in the ipsilateral MLF
- Accommodation intact
- Multiple sclerosis
- Other pontine lesions such as CVA
Acute 1½ Syndrome
(Paralytic Pontine Exotropia)
Where pathway

What pathway

Cortical Vision Disorders

- Prosopagnosia
  - Inability to recognize familiar faces
  - Right occipitotemporal lobe

- Balint syndrome
  - Triad of:
    - Simultagnosia
    - Optic ataxia
    - Visual apraxia
  - Bilateral parietal lobe lesions
References/Resources

- www.eyeweb.org
- emedicine.medscape.com/ophthalmology
- umed.med.utah.edu

- Basics in neuro-ophthalmology by Kathleen B. Digre, MD
- Using the ophthalmoscope and viewing the optic disc by Kathleen B. Digre, MD

- www.jeffmann.net/Neuroguidemaps
- uuhsc.utah.edu
- www.kellogg.umich.edu
- www.richmondeye.com
Questions and Answers