Neuro-ophthalmology

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Neuro-ophthalmology

• Study integrating ophthalmology and neurology
• Disorders affecting parts of CNS devoted to vision or eye:
  • Afferent system (visual pathway, incl. optic nerve)
  • Efferent system (ocular motor control, pupillary function)
Part I

Neuro-ophthalmologic Examination
Examination

• History
• Eye examination (visual acuity, tonometry, anterior segment examination, funduscopic examination)
• Perimetry
• Color vision, contrast sensitivity, electrophysiology (ERG, VEP)
• MRI of brain,
• Neurologic examination
Visual acuity

• Each eye separately
• Distance and near vision
• Using of corrective lenses, pinhole
• Using Snellen chart (20 feet) – normal 20/20
• Count fingers, hand motion, light perception, no light perception
Color vision

- Each eye separately
- Comparison between eyes
- Examination:
  - pseudoisochromatic plates (Ishihara)
  - 100 Hue test (Farnsworth-Munsell)
Farnsworth-Munsell 100 Hue test

- Ordering the color tiles as patient sees it
Contrast sensitivity

- Examining spatial frequency
- Decreased in some optic nerve disorders (typically optic neuritis)
Perimetry

• To assess the quality of visual field
• Characteristic visual field defect = location of possible intracranial lesions
Perimetry

- Automated static perimetry
Perimetry

- Goldmann kinetic perimetry
Electrophysiologic examination

ERG = Electrotretinography

- Access possible functional pathology of retina (scotopic, photopic and central part)
- **Flash ERG** (activity of bipolar cells as an answer to stimulation of photosensitive cells – rods, cones)
- **Pattern ERG** (activity of ganglionar cell as a response to stimulation of cones in macula)

VEP = Visual evoked potentials (responses)

- Access the capability of anterior visual pathways – optic nerve
- Major use: diagnosis/confirm of optic neuritis
Electrophysiologic examination
Electroretinography

Normal

Rod Specific
- Rod Function
  - b-wave

Maximum Scotopic
- Mixed Rod and Cone Function
  - b-wave
  - a-wave

Photopic 30 Hz Flicker
- Cone Function
  - b-wave

Transient Photopic
- Cone Function
  - b-wave
Visual evoked potentials

Pattern-Reversal VEP
15' checks, 3.8 reversals/sec

Latency
Normal

Latency
Delayed

102.5 ms
134.7 ms

32.2 ms

OD
OS
Multifocal ERG, Multifocal VEP

- Mostly experimental use, not standard in clinical medical practice here

![Normal responses vs. Stargardt’s disease in multifocal ERG and VEP](image)
Part II

Pathology of Afferent system
Afferent system

- **Retina** (cones, rods, bipolar and ganglion cells)
- **Optic nerve**
- **Optic chiasm**
- **Optic tract**
- **Lateral geniculate body**
- **Optic radiation**
- **Visual cortex** ($V_1$ = Brodmann area 17)
Pathologies of Afferent Visual System

• Papilledema

• Optic Neuritis

• Optic Neuropathy

• Optic Atrophy
Papilledema

• Not a disease - sing secondary due to elevated intracranial pressure (ICP)
• Unspecific sign
• Require immediate diagnosis = increased ICP is a life-threatening situation!!!
• 60% of cases = increased ICP caused by intracranial tumor!!!
• Other possible causes: hydrocephalus, meningitis, encephalitis, brain abscess...
Papilledema

Clinical picture

Early
• Margins are obscured
• Optic cup initially preserved
• Hyperemic disc

Acute
• Elevation of disc
• Radial hemorrhages
• Grayish-white exudates

Chronic
• Disc edema
• Obiterated optic cup
Optic neuritis

• Inflammation of the optic nerve
• **Intraocular** – within the globe
• **Retrobulbar** – posterior to the globe
• Usually unilateral
• Tendency to repeat

**Etiology**

• Often associated with multiple sclerosis (MS) = demyelinating optic neuritis (20% = first sign of MS)
• Other possible inflammatory causes: Lyme disease, syphilis, inflammation from orbit, paranasal sinuses...
Optic neuritis

**Symptoms**
- Sudden vision loss within several hours (mild blurring/light perception)
- Central, paracentral scotoma
- Retrobulbar/parabulbar pain
- Present afferent pupillary defect

**Prognosis**
- depends on underlying disorders
- MS = usually good – significant spontaneous improvement (several weeks)
- Some permanent disturbances of vision are possible (color vision decreasing, scotoma)
**Anterior Ischemic Optic Neuropathy**

**Etiology**
- Acute disruption of blood supply (due to vascular changes, infarction)

**Symptoms**
- Sudden unilateral loss of vision
- Altitudinal or wedge-shaped visual field defect
- Present afferent pupillary defect

**Clinical picture**
- Edema of optic disc
- Segmental obscuration of margins (correlation with visual field defect)
Anterior ischemic optic neuropathy

- 2 forms
- Benign: Nonarteritic AION
- Malign: Arteritic AION

Arteritic AION

- Association with systemic vasculitis (giant cell arteritis)
- Diagnosis: sedimentation rate, biopsy of temporal artery
- High risk of affection of contralateral (fellow) eye within days/weeks!!!
- Need for immediate therapy with high dose intravenous corticoids!!!
# AION forms

<table>
<thead>
<tr>
<th></th>
<th>Arteritic form</th>
<th>Non-arteritic form</th>
</tr>
</thead>
<tbody>
<tr>
<td>% of cases AION</td>
<td>10 %</td>
<td>90%</td>
</tr>
<tr>
<td>age</td>
<td>70 years</td>
<td>60 years</td>
</tr>
<tr>
<td>Sex</td>
<td>Female &gt; male</td>
<td>Female = male</td>
</tr>
<tr>
<td>Systemic disease</td>
<td>Giant cell arteritis (Horton disease)</td>
<td>idiopathic</td>
</tr>
<tr>
<td>association</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Prognosis</td>
<td>Very rare</td>
<td>mild</td>
</tr>
<tr>
<td>Fellow eye affection</td>
<td>often (50-90%)</td>
<td>rare (10-20%)</td>
</tr>
<tr>
<td>Diagnostics:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sedimentation (FW)</td>
<td>Very high</td>
<td>normal</td>
</tr>
<tr>
<td>treatment</td>
<td>High dosage of systemic corticoids</td>
<td>Not available</td>
</tr>
</tbody>
</table>
Optic Atrophy

• Irreversible loss of axons as a result to damage of optic nerve

Etiology

• **Primary** due to trauma, direct pressure by tumor
• **Secondary** due to affection of optic nerve (optic neuritis...)
• **Glaucomatous** due to glaucomatous damage

Pathogenesis

• **Ascending** - lesion located anterior to the lamina cribrosa
• **Descending** – lesion located posterior to the lamina cribrosa
Optic Atrophy

Clinical picture
• Total/partial pale optic disc
• Well defined / blurred margins
• Constricted / reduced retinal vessels

Etiology
• Vascular (AION, RAO)
• Inflammation (optic neuritis, neuroinfections)
• Compressive (orbital/intracranial mass)
• Traumatic (avulsion, bone fracture)
• Toxic (methyl alcohol, various poisons, cytostatics)
• Congenital/hereditary (LHON, Kjer atrophy)
• Systemic (hematooncological diseases)
Part III

Pathology of Efferent system
Efferent system

• 1) Cranial neuropathies (III, IV, VI)

• 2) Pupillary abnormalities
Eye movement

- Ocular motility – produced by extraocular muscles
- 4 rectus muscles (lateral, medial, superior, inferior)
- 2 oblique muscles (superior, inferior)
Cranial neuropathies

**Signs**

**Oculomotor nerve palsy**
- Diplopia
- Multiple muscle paralysis
- Ptosis
- Anisocoria

**Trochlear nerve palsy**
- Vertical diplopia
- Abnormal head tilt

**Abducens nerve palsy**
- Horizontal diplopia in the gaze palsy
Cranial neuropathies

Etiology

- Ischemic (diabetes, hypertension, hyperlipidemia)
- Demyelinating disease (MS)
- Compressive (tumor, aneurysm)
- Elevated ICP

- Multiple cranial neuropathies = suspect lesion in the posterior orbit or cavenrous sinus region
Pupil

- **Miosis** – parasympathetic nervous system
- **Mydriasis** – sympathetic nervous system
Sympathetic pathway
Pupillary abnormalities

Anisocoria

• inequality of pupil size
• May be physiologic
• Possible accidental discovery
• May be isolated / associated with eyelid or ocular motility abnormalities

Diagnosis

• Direct shine at pupil
• Test near response (miosis with accommodation)
• Pupil sizes in light and dark
Horner’s Syndrome

**Signs**

- Miosis (pupil does not dilate in dark)
- Ptosis
- Pseudo-enophthalmus
- Anhidrosis (diminished sweating)
- Heterochromia (if congenital)

**Etiology**

- Trauma, internal carotid artery dissection, brain stem strokes, MS, brain tumor, syringomyelia, apical lung tumor, goiter, thyroid carcinoma...
Adie’s Pupil

**Signs**
- No present / slow miosis to light
- Present miosis to accommodation
- Pupil is larger with light/near dissociation

**Etiology**
- Inflammation (viral or bacterial infection)

**Therapy**
- Pilocarpine drops, thoracic sympathectomy
Thank you for your attention!