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JCAHPO Regional Meetings
2017
You have some Nerve Asking Me to Work Up that Patient!

What I Need to know about the Neuro-Ophthalmology Patient

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What is a Neuro-Ophthalmologist?
• "Neuro-ophthalmologists take care of visual problems that are related to the nervous system; that is, visual problems that do not come from the eyes themselves.

North American Neuro-Ophthalmology Society

What is a Neuro-Ophthalmologist?
• We use almost half of the brain for vision-related activities, including sight and moving the eyes."

North American Neuro-Ophthalmology Society

What is a Neuro-Ophthalmologist?
• "Neuro-ophthalmology, a subspecialty of both neurology and ophthalmology, requires specialized training and expertise in problems of the eye, brain, nerves and muscles.

• Neuro-ophthalmologists complete at least 5 years of clinical training after medical school and are usually board certified in Neurology, Ophthalmology, or both."

North American Neuro-ophthalmology Society

What is a Neuro-Ophthalmologist?
Some of the common problems evaluated by neuro-ophthalmologists include:
• optic nerve problems (such as optic neuritis and ischemic optic neuropathy)
• visual field loss
• unexplained visual loss
• transient visual loss, visual disturbances
• double vision
• abnormal eye movements
• thyroid eye disease
• myasthenia gravis
• unequal pupil size
• eyelid abnormalities."

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Components of the Neuro-Ophthalmic Exam

- History
- Review of Systems and Other History
- Visual Acuity
- Amsler Grid
- Color Vision
- Brightness Sense Testing
- Confrontation Visual Fields
- Tests of Stereopsis
- Motility and Alignment Exam
- Pupillary Exam

Components of the Neuro-Ophthalmic Exam (continued)

- External Exam
- Slit Lamp Exam and Tonometry
- Automated or Goldmann Perimetry
- Ophthalmoscopic Exam
- Neuroimaging
  - Computed tomography
  - Magnetic resonance imaging
  - Imaging of the orbit
  - Imaging of the brain
  - Cerebral angiography

History

- Visual complaints are often difficult to articulate
- Patients usually have an array of disjointed complaints
- Even if the history reveals something of more serious concern, DO NOT IGNORE the Chief Complaint

Details of the patient's history are important and often determine the diagnosis

Visual Complaints

- Loss of Vision
- Double Vision
- Eye Pain
- Negative or Positive Visual Phenomenon

Loss of Vision

- Monocular or Binocular
- Transient or Permanent
- Tempo of Visual Loss
- History of Past Episodes
- Any other associated symptoms

Eye Pain

- Location of Pain (periocular, retro-ocular, etc)
- Transient or Permanent
- Frequency and Severity
- Any other associated symptoms
Positive Visual Phenomenon
• Photopsias, phosphenes
• Moore’s lightning streaks
• Scintillating scotoma
• Unformed hallucinations
• Formed hallucinations
• Floaters
• Afterimages

Negative Visual Phenomenom
• Blindness
• Blurriness
• Dim
• “A skim”
• “A cloud”
• “A curtain”
• Screen covering vision
• Washed out color

Double Vision
• Monocular or binocular
• Transient or permanent
• Tempo of onset
• History of diplopia in past

Visual Acuity
• Snellen eye chart (distance)
• Near vision card
• Picture chart
• Tumbling E chart
• Low Vision Tests:
  • Counting-fingers acuity (CF or FC)
  • Hand-movements acuity (HM)
  • Light-perception acuity (LP or NLP)
Amsler Grid
- Square containing small grid lines with a central target
- Most useful in identifying metamorphopsia (a disturbance of vision in which objects are seen as distorted in shape) i.e. macular disease

Amsler Grid
- Also useful test of central visual field
- At standard reading distance, the grid covers about 10 degrees of visual field in each direction from fixation
- Central visual field defects may make it difficult for patients to see the central dot, or they may note that some of the squares or lines are missing or are lighter than others

Color Vision
- Acquired color vision deficits may be caused by optic nerve or retinal disease
- Optic nerve disorders tend to affect red green discrimination
- Retinal disorders create more blue/yellow confusion

Color Vision
- Numbers are hidden in a matrix of seemingly random dots
- These color plates originally designed for congenital color deficits
- Yet, total number of plates missed is often helpful to determine acquired color vision deficits
**Stereoacuity Testing**
- Measured viewing with polarized glasses
- It is the brain processing 2 disparate retinal images

**Ocular Motility**
- Check for abnormalities in eye movements and for ocular alignment
- 3 procedures:
  - 1. Measure ocular movements
  - 2. Corneal light reflection test
  - 3. Cover test

**Ocular Movements**
- Observe for any rhythmic oscillations (nystagmus)

**Corneal Light Reflection Test**
- Corneal light reflex tests useful in assessing ocular alignment for those not able to cooperate with cover testing or who have poor fixation
- Clinical examples of this Testing Type:
  - Hirschberg test
  - Krimsky test

**Cover Test**
- More accurate test for ocular misalignment
- Requires more cooperation from the patient and more skill on part of the examiner
**Visual Field Tests**
- Confrontation Visual Fields
  - Performed one eye at a time
  - Test the ability of the patient to count fingers in the four quadrants of the visual field

**Visual Field Homonymous Hemianopia**
- Homonymous hemianopia implies loss in one of the half-fields; loss of either the right or left half of the visual field in either eye

**Pupillary Exam**
- 3 components to exam:
  1. Measure pupil size in dim light (assesses the motor, or efferent, limb of the reflex arc)
  2. Evaluate pupil response to direct light (assesses both the motor and sensory, or afferent, limbs)
  3. Swinging-light test (assesses only the sensory limb)

**Clinical Assessment of Pupils**
- Pupils that differ in size (ANISOCORIA) by more than 0.5-1mm in dim light may be clinically important
- Anisocoria can be obscured if the pupils are constricted, therefore best to perform in the dimmest light possible
Objectives

Direct Ophthalmoscopy
- Produces an upright, or unreversed image of approximately 15x magnification of the retinal fundus

Here comes your first patient…..

Case
- 26 y/o female BMI = 32.0
- Inc HA x 3months
- Bl VA OU intermittently
- TVOs OU more recently

Case
- ROS – non-contributory
- No associated conditions
- Vision OD 20/15 OS 20/15-3
- Ishihara OU 11/11
- Amsler Grid no metamorphopsia
- Stereo 50 sec of arc

Case
- No desaturation to color red
- Subjective brightness equal OU
- EOM – full ductions OU; no misalignment
- Pupils- NO RAPD
Idiopathic Intracranial Hypertension (IIH)

- Also called Pseudotumor Cerebri (PTC)
- Disorder of inc ICP of unknown cause
- Incidence 22.5 new cases each year per 100,000 overweight females of childbearing age
- Rare, males

Criteria for Diagnosis of IIH

- Signs & Symptoms of increased ICP or papilledema
- Normal neuroimaging
- Elevated ICP by LP (lateral decubitus position)
- Normal CSF composition
- No other cause of inc ICP identified

Case

- MRI Brain/Orbit study – nl
- MRV Brain – nl
- LP OP = 31 cm H₂O

Case – Medical Therapy

- Start Diamox 500mg TID
- Stress low sodium diet
- Exercise
- Weight loss

Humphrey Visual Fields
Enlarged Blind Spots OU
Idiopathic Intracranial Hypertension (IIH)

- Most suffer debilitating HA
  - Usually severe, daily, and often, throbbing
- Pulsatile intracranial noise or pulse-synchronous tinnitus
  - Often unilateral
- Yet, the most serious problem is vision loss

Idiopathic Intracranial Hypertension (IIH) TVOs

- Transient visual obscurations (TVO)
  - Occur in 75% of IIH pts
  - May involve one or both eyes
  - Not associated with poor visual outcome

IIH and Visual Loss

- 86% visual loss
- 10% severe visual loss
- Approx 5% blind (or at least in one eye)

Treatment for IIH
Medical vs Surgical

- Weight Loss, Diet
- Diuretics (Acetazolamide, Lasix)
- Repeated spinal taps
- ONSF
- CSF shunting procedures (VP or LP shunt)
CC: “I suddenly lost vision in one eye”

Case

ROS
• No preceding viral illness

PMHX
• None

PSHX
• No TOB, ETOH

FHX
• Non-contributory

Examination

• VA OD 20/20 OS 20/80
• Ishihara OD 11/11 OS 04/11

• Desaturation to color red OS
• Decrease subjective brightness OS
• Stereo 80 seconds of arc

Fundus

Slit Lamp Examination
Anterior Segment – normal OU
IOP- 15 mm Hg OU
Optic Neuritis

• Acute inflammatory disorder of the optic nerve
• Commonly associated with Multiple Sclerosis

Optic Neuritis

• Acute, unilateral visual loss
• Pain with eye movements
• 65% with normal appearing optic nerve

So, What's the Concern of your Patient?

• Optic neuritis may be the first clinical sign of MS

• MRI Brain/Orbit imaging study with gadolinium should be considered in all patients with optic neuritis to assess the risk to MS.

• In the absence of demyelinating lesions, the risk is low at 25% while the risk is 72% when lesions were present.

How to treat Optic Neuritis

• The clinical decision to manage and treat a patient with typical optic neuritis with steroids is often challenging.

• In summary, the ONTT revealed the IV steroid treated patients recovered visual function faster within the first 4-6 weeks post onset of optic neuritis, but showed no statistical difference in final visual outcome between the IV steroid treated patients and those receiving placebo.
How to treat Optic Neuritis

- Oral steroids are contraindicated for acute optic neuritis because of a higher rate of recurrence of optic neuritis.

- But all patients with abnormal MRIs should be referred to the neurologist for further evaluation and management for MS.

Oh, My Technician…..Please, can you work up one more patient today?

What you don’t want to Miss!

Case

- 79 y/o female with h/o dull, retro-ocular pain for 2 wks prior and a new “light” HA

- Sudden loss of vision OD x 2 days

Case

- POH - s/p cat ext OU 1993; ARMD (dry form)
- Ocular Rx - Ocudyne
- PMHX - non-contributory
- Systemic Rx - Premarin, Provera, Bilberry
- NKDA
- SoHx - Divorced, 1 daughter, no tob

Case

- ROS:
  - “light” HA not sure if in temporal area
  - No scalp or temporal tenderness
  - No recent weight loss or loss of appetite
  - No jaw claudication
  - No fever
  - No joint pain
  - No fatigue

Case

- Va : FC OD 20/25 OS
- Color (Ishihara) Plates: 02/11 OD 11/11 OS
- Amsler Grid: no metamorphopsia
- Desaturation to color red OD
- Pupils: 5.0mm OU
  - >1.8 log unit Right RAPD
  - HVF
Case
- EOM - full ocular ductions OU, no misalignment, no nystagmus
- Slit lamp biomicroscopic exam: bilateral pseudophakia
- IOP: 15 mmHg OD  12 mmHg OS

Fundus Right Eye: Pallid Disc Edema

Case
- BP = 196/72
- ESR = 117
- C-reactive protein = 2.7

Case
- Diagnosis: Arteritic Ischemic Optic Neuropathy
- Admitted for High Dose IV Steroids and Temporal Artery Biopsy

Giant Cell Arteritis
Sight-threatening,
Systemic vasculitis
Giant Cell Arteritis
High index of suspicion = Prompt diagnosis & Expeditious Treatment

Clinical Manifestations of GCA
Headache
- aches over the temporal or occipital areas, unilateral or bilateral
- described as piercing or associated with lancinating pain

Clinical Manifestations of GCA
• discomfort over the skin of the scalp or face or anywhere along the distribution of the occipital, superior temporal, facial, or lingual arteries

• Jaw Claudication is a classic sign—— considered to be pathognomic

Clinical Manifestations of GCA
• malaise
• migratory arthralgias
• proximal muscle pain and stiffness

Clinical Manifestations of GCA
• flu-like symptoms
• anorexia
• weight loss
• low grade fever
• night sweats
• weakness
• depression

Clinical Manifestations of GCA
• amaurosis fugax
• diplopia
• sudden loss of vision
Giant Cell Arteritis

- Prompt treatment with high-dose steroids may prevent further unilateral or bilateral visual loss from anterior or posterior ION or CRAO

Now you have the NERVE to take care of any neuro-ophthalmic patient!

Thank you

Drawn by the kindergarten students at Episcopal, Baton Rouge
October 2011 “Me & My Service Eye Dog”
Visit to discuss “How the Eye Works”