NEURO-OPHTHALMOLOGICAL EMERGENCIES PRESENTING IN OPTOMETRY

Jacqueline Theis, OD, FAAO
OptoWest

Goals/General Outline
- Outline the signs, symptoms, and optometric management of neuro-ophthalmological emergencies
- Identify questions providers should add to their patient history
- Describe the epidemiology, ocular and systemic manifestations, diagnosis and optometric management of:
  - Giant Cell Arteritis
  - Horner’s Syndrome
  - Intracranial (Posterior-communicating artery) Aneurysms
  - Myasthenia Gravis
  - Intracranial Space Occupying Lesions
    - Cavernous Sinus Lesions
    - Pituitary Apoplexy

Disclosure Statement:
C. Light Technologies – Clinical Research Consultant

“Neuro-ophthalmological emergencies constitute vision or life-threatening conditions if diagnosis and treatment are not promptly undertaken.

Even with immediate therapy, these clinical entities carry a high rate of morbidity.”


Symptoms of Neuro-Ophthalmological Emergencies
- Vision Loss
- Diplopia
- Eye Pain
- Headache

PEARL for Concern: If you have more than one of the following
- Pupil abnormality
- Eyelid abnormality
- EOM abnormality


Signs of Neuro-Ophthalmological Emergencies
- Optic nerve edema or pallor
- Extraocular/intraocular abnormality
- Multiple cranial nerve palsies
- Pupil-involving CN III Palsy
- Anisocoria
- Ptosis
Patient History in a Neuro-Ophthalmological Emergency

<table>
<thead>
<tr>
<th>Location</th>
<th>Monocular or Binocular?</th>
</tr>
</thead>
<tbody>
<tr>
<td>Extent</td>
<td>Gaze dependent?</td>
</tr>
<tr>
<td>- Left vs. Right, Up vs. Down</td>
<td>Central or peripheral visual field?</td>
</tr>
<tr>
<td>Distance vs. Near</td>
<td>Left vs. right visual field?</td>
</tr>
<tr>
<td>Onset</td>
<td>When did it start?</td>
</tr>
<tr>
<td>- Sudden or gradual?</td>
<td>What were you doing?</td>
</tr>
<tr>
<td>Frequency</td>
<td>Is it getting better, worse or staying the same since it started?</td>
</tr>
<tr>
<td>Duration</td>
<td>How long does it last?</td>
</tr>
<tr>
<td>- Intermittent or constant</td>
<td>(seconds, minutes, hours or days)</td>
</tr>
<tr>
<td>Timing</td>
<td>Is it worse at the beginning or end of the day?</td>
</tr>
<tr>
<td>What has it happened before?</td>
<td>History of childhood strabismus, previous eye surgery?</td>
</tr>
</tbody>
</table>

Monocular vs. Binocular Diplopia

1) Do you see double with both eyes open? 
   - Yes
   - No

2) Cover the right eye, do you still see double? 
   - Yes (Monocular)
   - No

3) Cover the left eye, do you still see double? 
   - Yes
   - No (Binocular)

Examination

- Distance Visual Acuity
- Pinhole Acuity
- Color Vision
- Ishihara, ACHR, Red Cap
- Pupils
  - Size in light/dark
  - Reaction to light/dark
  - Near response
  - RAPD?
- Visual Fields
  - Confrontation
  - Automated
- Eyelids (MRD1/MRD2)
- Optic Neuropathy
- Fundus Evaluation
- Cranial Nerve Evaluation

Acute Vision Loss

1) Does visual acuity improve with a pinhole? 
   - Yes
   - No

2) Pupils - Presence of RAPD? 
   - Yes
   - No

3) Visual Field Defect? 
   - Central
   - Peripheral
   - Constriction, Enlarged blind spot

- Optic disc pallor
- Chiasm lesion – pituitary adenoma
- MRI/Brain contrast
- MRA/MRV, Lumbar puncture

Giant Cell Arteritis

Epidemiology

**GCA**
- Most common systemic vasculitis affecting adults >50y
- Rare in people <50y
- Average age of onset is 74-76y
- For each decade after 50, incidence increases from 2.0 (50-60y)
- 11.8 (61-70y)
- 31.3 (71-81y)
- Women affected 2-3x more than men
- More common in whites, Nordic/Northern European ancestry, and other northern latitudes

**AAION**
- Annual incidence of AAION from GCA is 1.3 per 100,000
- Vision loss from AAION and CRAO from GCA is severe
- 73% present with VA worse than 20/200
- 15% of eyes have an improvement, likely from eccentric fixation

Pathogenesis

**Persistent vessel wall inflammation + vascular damage + stenosis, occlusions, and aneurysms**

**why is it an emergency?**
- **Blindness**
- **Vision loss ~10% of patients**
- **Stroke**
- **Aortic aneurysm or dissection**

**Systemic Manifestations**

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<thead>
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<th>Symptoms</th>
<th>Signs</th>
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<td>Sudden visual loss</td>
<td>AAION (6.9%) – 1.3 per 100,000 population</td>
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<tr>
<td>Most frequent symptom ~50% of cases</td>
<td>(+) RAPD</td>
</tr>
<tr>
<td>Transient visual loss ~30%</td>
<td>(±) Pseudo exudation</td>
</tr>
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<td>Often followed by permanent visual loss</td>
<td>Accounts for 85% of cases of permanent vision loss</td>
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<tr>
<td>Diplopia ~6%</td>
<td>CRAO (1.6%)</td>
</tr>
<tr>
<td>Eye pain ~8%</td>
<td>CilioRAO (0.4%)</td>
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<td>Posterior ION</td>
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Ocular Manifestations

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Pathogenesis

**Infectious? immune trigger in a genetically predisposed subject**
- T-cell mediated granulomatous inflammation of medium- and large-vessels
- Aorta
- External carotid artery
- Posterior ciliary artery
- **Posterior temporal arteries**

**Why is it an emergency?**
- The sooner GCA is diagnosed and treated, the lower the incidence of visual loss
- However, patients presenting with poor vision have little chance of recovery despite immediate steroid treatment
- The main goal of treatment is to prevent vision loss in the fellow eye
- Usually occurs within days in 50% of cases of untreated GCA

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<td>Jaw Claudication (48%)</td>
<td>20% of cases with permanent vision loss from GCA may present without systemic symptoms of GCA</td>
</tr>
<tr>
<td>Neck pain (17%)</td>
<td>Temporal artery tortuosity, prominence, and/or tenderness</td>
</tr>
<tr>
<td>Headache (57%)</td>
<td></td>
</tr>
<tr>
<td>Scalp tenderness (20%)</td>
<td></td>
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<tr>
<td>Weight loss (40%)</td>
<td></td>
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<tr>
<td>Anorexia (31%)</td>
<td></td>
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<tr>
<td>Myalgias (28%)</td>
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<td>Malaise (37%)</td>
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## Diagnosis

**Serological Studies**
- Elevated ESR (85.7% sensitivity)
- Normal in 9.2-14.3%
- Elevated C-Reactive Protein (97.5% sensitivity)
- Normal in 1.7%
- ESR/CRP can also be elevated in infection and cancer
- Abnormal CBC w/ differential (sensitivity ≤86%)
- Thrombocytosis
- Normocytic anemia
- Leukocytosis

### Fluorescein angiography
- Differentiate AION from NAION
- choroidal hypoperfusion
- Delayed choroidal filling

### Temporal Artery Biopsy
- Gold Standard
- BUT only 60-85% of patients with GCA have a (+) TAB
- May need sequential if high clinical suspicion

**Horner’s Syndrome**

1. **MYDRIASIS**
   - Postganglionic neuron travels from sympathetic chain to cavernous sinus, then to internal carotid artery
   - Acts on carotid sinus to increase parasympathetic tone

2. **PUPIL PATHWAY REVIEW**
   - Efferent (3 neuron chain)
     - Miotic (3rd neuron)
     - Iris dilator axon enters the ciliary ganglion

   - **Abnormal:** Pupil is larger in the dark

## Differential Diagnosis

- **Common or migraine headache**
- **Atherosclerosis of large vessels**
- **NA-AION**
- **Takayasu arteritis**
- **Other forms of vasculitis**
- **Polyarteritis nodosa**
- **PMR**
- **Multifocal motor neuropathy**
- **Thyroid eye disease**
- **Other forms of vasculitis**

## Treatment

- **Steroids** – IMMEDIATELY once AION is suspected
  - Oral (80mg po) vs. IV ASAP
  - Oral taper over months to year
  - Low dose Aspirin?
  - Consult PCP/internist
  - Monitor for steroid-related complications – hypertension, diabetes, osteoporosis, infection, etc.
  - Smoking cessation
  - Follow up – 2-4 weeks

- Only 4% of patients will improve visual loss with steroids
- 4% of patients lose vision within the first 5 days, even on steroid treatment

**Future**

- **Glucocorticoids are very effective at high doses**
  - Relapses occur in up to 50% of patients when doses are tapered
  - Not ideal for chronic management
- **Adjunct immunosuppressants**
  - Methotrexate
  - Reduce risk of relapses
  - Positive effects may take 6-8 months to emerge
  - Anti-TNFα (Infliximab, Adalimumab, Etanercept)
  - No additional benefit above prednisone monotherapy
  - Increased risk of infection
  - Anti-IL-6 (Tocilizumab)
  - May provide additional benefit to prednisone by inducing and maintaining remission for up to 52 weeks. Quick onset
  - Other targeted anti-inflammatory therapies
  - Abatacept – reduced risk of relapse
  - Ustekinumab – glucocorticoid sparing response
  - Rituximab

**Polymyalgia rheumatica**

- PMR is 2-3x more common than GCA

## Pupil Pathway Review: Sympathetic

- **Efferent (3 neuron chain)**
  1. Hypothalamus–pituitary center of Bulge at C8-T2 (spinal cord)
  2. Through sympathetic chain (adjacent lung apex) → superior cervical ganglion (located at level of carotid bifurcation)
  3. Postganglionic neuron travels with the internal carotid artery until cavernous sinus, then follows C1N→CNV1 – long ciliary nerves → iris dilator → MYDRIASIS

**Abnormal:** Pupil is larger in the dark.
**Epidemiology of Carotid Artery Dissection**

**Epidemiology**
- Incidence of 2.6-5 per 100,000 population
- Peak incidence – 5th decade
- Mild male predominance
- Responsible for 25% of strokes in young adults (<45yo)

**Ocular Manifestations of Horner’s Syndrome in Carotid Artery Dissection**

- Painful third-order Horner Syndrome (~60%)
  - Ptosis (~1-2mm)
  - “Inverse ptosis” – when lower lid is slightly elevated
  - Miosis (anisocoria of 1-1.5mm)
  - Dilation Lag
- WITHOUT facial anhidrosis or partial
- Less common ocular signs of CAD
  - Transient monocular vision loss
  - NAION
  - CRAO
  - Ocular ischemic syndrome
  - Ocular motor nerve palsies

**Systemic Manifestations**
- Ipsilateral headache (~70%)
- Ipsilateral neck pain (10-30%)
- Ipsilateral ear pain (10-30%)
- Less common
  - Lower cranial nerves affected
  - Pulsatile tinnitus
  - Vertigo
  - Dysgeusia (foul taste in the mouth)
  - Headache ~70%
  - Ear pain (10-30%)
  - Neck pain (10-30%)
- Diseases that affect the brainstem, spinal cord, chest or neck can present with purely ocular symptoms
- Ophthalmologic signs/symptoms can proceed ocular or cerebral infarction in 33% of patients 6-14 days

**Diagnosis**

**Homer’s Syndrome**
- Pharmacologic diagnosis/localization
  - (+) = Failure of pupillary dilation after one hour (ie anisocoria >1mm remains)
  - Apresartanline 1%
  - (+) = mydriasis of affected pupil >1mm 30-45 minutes after drop instillation
  - Hydroxyamphetamine
  - Localizes between central (1st)/
    preganglionic (2nd) and postganglionic (3rd) order lesion
  - If mydriasis >1½2/4 order
  - (-) mydriasis (ie anisocoria 1mm remains)→ 3rd order
  - Caucets
  - Can perform cocaine and hydroxyamphetamine test on the same day (within 24-48 hours)
  - Hydroxyamphetamine test may yield a false-negative, and is not commercially available

**Carotid Artery Dissection**
- MR/MRA
  - 85% sensitivity
- CTA of head/neck or carotid doppler ultrasound
- CT of chest
- CBC with differential
Intracranial Aneurysms

Epidemiology
- 9-36% of CNIII palsies are caused by an intracranial aneurysm
- Posterior communicating artery (PCA) aneurysms present with a CNIII palsy 30-60% of the time
- 40% of aneurysms are located at the level of the PCA, ophthalmic artery, and cavernous sinus

Risk factors for Aneurysmal Rupture
- Increasing age (peak 6th decade)
- Female gender
- Smoking
- Hypertension
- Heavy alcohol consumption
- (+) Family history of intracranial aneurysm or subarachnoid hemorrhage
- Aneurysm size >10mm
- Genetic disorders – polycystic kidney disease
- Aneurysm location
  - PCA and basilar tip have higher rupture risk at 5 years

Ocular Manifestations
- Pain
- Mid-dilated pupil
- Poor or absent light reaction
- Complete or partial external CNIII palsy

Rule of the Pupil
- Complete CN III
  - Pupil-sparing – likely ischemic
  - Pupil-involving – likely compressive
- Incomplete CN III
  - Pupil-sparing – ????
    - 14% will have aneurysm, just present to office in an early phase before the pupillary fibers are involved
  - Pupil-Involving – compressive

Topographical localization of CNIII Palsy
- Brainstem
- Hemiparesis
- Hemisensory loss
- Other cranial neuropathies
- Subarachnoid space
  - Meningeal signs
  - Stiff neck
  - Severe headache
- Other cranial neuropathies
- Cavernous sinus
  - CN – 4, 5, or 6 involvement
  - Horner syndrome
- Orbit
  - Proptosis
  - Chemosis
  - Optic neuropathy
Diagnosis and Management

Diagnosis
- Digital subtraction angiography (DSA)
- 1-2% morbidity risk
- Gold standard
- MRA/CTA
- Noninvasive
- Can detect 95% of aneurysms
- Aneurysm needs to be >5mm
- MRI/CT/LP
- ESR/CRP/CBC w/ differential

Management
- 70-100% of surviving patients make a complete or partial recovery of the oculomotor deficit
- Usually starting with resolution of ptosis
- Pupillary and EOM abnormalities may persist
- Sometimes aberrant regeneration
- Fresnel prism please


Myasthenia Gravis

Epidemiology
- Incidence – 0.4-5 cases/100,000 per year
- Prevalence – 0.5-30 cases/100,000 per year
- Age
  - Juvenile MG
    - 10-15% of Caucasians (28)
  - 50% of Chinese cases (29)
  - Early-Onset MG (EOMG) <50yo
    - Female predominance (60-70%)
  - Between 50-60yo
    - No gender difference
  - Late-Onset MG (LOMG) >60yo
    - Male predominance


Pathophysiology
- Neuromuscular junction dysfunction leads to painless, fatigable weakness of voluntary muscles
- Autoantibodies
  - Anti-AChR
  - Anti-MuSK
- Role of the Thymus?
  - Pathogenic/inflammatory triggers→chronically inflammed thymus→autosensitization to AChR→autoreactive T Cells

Ocular Manifestations
- Ptosis
  - variable, worse with fatigue
- Diplopia/EOM involvement/ Ophthalmoplegia
  - variable, worse with fatigue
- Orbicularis weakness
- Normal pupils
- Cogan’s Lid Twitch


Systemic manifestations
- 85% of MG patients
- Skeletal Muscle Fatigue
  - Facial muscles
  - Proximal limb muscles
  - Muscles for swallowing/breathing
  - Other autoimmune disease
    - Thyroid disorders (Hashimoto’s or Basedow diseases)
    - Thymus problems
      - Thymic follicular hyperplasia ~70%
      - Thymoma (thymic epithelial cell tumor) ~10-15%

Image and information from:
Why is it life threatening?
- Myasthenic crisis
  - Respiratory failure due to muscle weakness
  - Severe weakness of respiratory muscles, upper airway muscles, or both
  - Usually due to poor control of generalized disease
- Other triggers for crisis
  - Concomitant use of certain antibiotic (aminoglycosides, quinolones, antimalarials), muscle relaxants, anti-inflammatory drugs, beta-blockers (including topical), and iodinated radiocontrast agents
  - Systemic infection involving respiratory tract, aspiration, and surgery
  - Emotional stress, hot environment, sudden elevation of body temperature
  - Hypothyroidism
  - Requires immediate ventilatory assistance
- Pre-immunotherapy era
- Myasthenic crisis had significant mortality rate (up to 75%), but has fallen to 5% in recent years
- 20-30% lifetime prevalence of myasthenic crisis in patients with MG
- Usually occurs during the course of first symptomatic presentation in the young and later in the course of disease in the elderly
- White patients more likely to respond poorly to treatment than black patients
- Pregnancy is known to aggravate MG

Serological Testing
- Generalized Myasthenia - 96% sensitivity, high PPV
  - 87% have autoantibodies
  - 85% AChR
  - 40-70% MuSK
- Ocular Myasthenia - 44% sensitivity, low NPV
  - 50-80% have autoantibodies
  - (+) Autoantibody assay highly suggests MG
  - BUT (-) Autoantibody assay = inconclusive
- Seronegative - (-) AChR, (-) MuSK
  - High prevalence of cranial and bulbar muscle involvement
  - Repeated serological tests
  - 15% seroconversion rate over 1 year period
  - Patients with OGM may persistently test seronegative

Diagnostic Management
- OD Diagnostic Tests - In Office
  - Ptosis \(\rightarrow\) Fatigue, Rest, Ice Tests
- OD/MD Diagnostic Tests (Neurologist/Neuro-Ophthalmologist)
  - Tension Test (Edrophonium Chloride), 95% sensitivity
  - Side Effects: Bradycardia, arrhythmia, hypotension
  - Serologic Testing for Autoantibodies, Thyroid function
    - 5% of patients with myasthenia gravis also have dysthyroidism
  - CT/MRI of Thorax (rule out Thymic abnormalities)
  - Neurophysiological Specific Tests
    - RNS - Repetitive Nerve Stimulation
    - SFEMG = Single Fiber Electromyography

Medical Treatment
- Long-Acting Acetylcholinesterase Inhibitor (Pyridostigmine)
  - Improves ptosis, less effective in resolution of EOM involvement/diplopia
  - Does not affect the course of the disease

Immunomodulatory Therapy
- Oral corticosteroids
  - Effectively controls diplopia AND ptosis symptoms
  - Lowers risk of progression from OGM\(\rightarrow\)GMG
- Other: Azathioprine, Mycophenolate Mofetil, Methotrexate, Tacrolimus, Cyclosporine, Rituximab
- Thymectomy
- IV Ig
- Plasma Exchange

Intracranial Space Occupying Lesions
- Tumor
- Inflammation
- Infection
- Ischemic infarct
- Increased intracranial pressure
- Mass
- Pseudotumor/IIH

Swollen Optic Nerve(s)
- Papilledema
- “Disc swelling from elevated intracranial pressure”

Differential Diagnosis
- Congenital Anomaly
- Optic disc drusen
- Infection
- Inflammation
- Ischemia

Until lumbar puncture - better to diagnose “optic nerve head edema”
Optic Nerve Edema – EVALUATION

1) SLOW YOURSELF DOWN AND BREATHE

- **Bilateral Disc Edema**
  - Initially superior-inferior swelling
  - Disc Hyperemia (its pink!)
  - (+) CWS/hemorrhages over time
  - (-) Spontaneous Venous Pulsation
  - Normal VA
  - Normal Color Vision
  - (+) Visual field defect
  - Enlarged blindspot
  - Peripheral field constriction

**Signs**

- Bilateral disc edema
- Initially superior-inferior swelling
- Disc Hyperemia (its pink!)
- (+) Obscuration of retinal vessels over disc margin
- Cup is preserved
- (+) CWS/hemorrhages over time
- (-) Spontaneous Venous Pulsation
- Normal VA
- Normal Color Vision
- (+) Visual field defect
- Enlarged blindspot
- Peripheral field constriction

**Diagnosis**

- Immediate Neuroimaging
- Lumbar puncture
- Elevated intracranial (CSF) pressure

**Etiology**

- Mass lesion
- Severe cerebral edema
- Venous thrombosis
- Hydrocephalus
- Pseudo-tumor Cerebri

**IIH - Management**

- Acetazolamide
  - May improve papilledema, visual complaints, headache
  - OR other diuretics/CAIs
- Weight loss
- Baseline automated VFs after treatment initiated
  - Progressive or severe vision loss may require more aggressive therapy
  - Ventriculopontine shunt
  - Optic nerve fenestration
Optic Disc Pallor

Longstanding compression of optic nerve, chiasm or tract by a tumor or aneurysm can cause progressive optic disc pallor

Cavernous Sinus Lesions

Cavernous Sinus Syndromes

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<th>Partial</th>
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<td>Ophthalmoplegia (Diplopia)</td>
<td>Depends on the location of the lesion</td>
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<td>Ptosis</td>
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<td>Mydriasis</td>
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<tr>
<td>Hypoesthesia of V1/V2 (facial numbness)</td>
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<td>Orbital Pain</td>
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Etiology

- Chronic Causes
  - Meningiomas
  - Metastases of head and neck cancers
  - Slow-growing carotid aneurysm
- Acute Causes
  - Cavernous sinus thrombosis
    - Extension of facial sinus infection
  - Carotid cavernous fistula
    - Painful red eye + chemosis + pulsatile exophthalmos
  - Inflammatory reaction (Tolosa-Hunt syndrome)
  - Pituitary apoplexy

Cavernous Sinus Thrombosis

Ocular Manifestations

- Most common 80-100% presentation
- Proptosis
- Chemosis
- Ptosis
- CN III, IV, and/or VI palsies
- Less common 50-80% presentation
- Periorbital edema
- Optic disc edema
- Venous engorgement
- Least common <50%
  - Decreased visual acuity (due to ION, CRADO, CRVO, or corneal ulceration)
  - Sluggish/dilated pupils
  - Periorbital and corneal sensory loss (CNV)

Systemic Manifestations

- Most common 80-100% presentation
- Acute onset fever
- Less common 50-80% presentation
- Headache
- Lethargy
- Altered sensorium
- Least common <50%
  - Meningismus
  - Seizures
  - Hemiparesis

Differential Diagnosis

Orbital Cellulitis

- Painful ophthalmoplegia
- Proptosis
- Chemosis
- Fever
- Decreased vision
- UNILATERAL

Direct High-Flow Carotid Cavernous Fistula

- Periorbital edema
- Ophthalmoplegia
- Increased IOP
- Decreased vision
- (+) supraorbital bruit
- Arterialized conjunctival vessels

Pituitary Apoplexy

Epidemiology
- Between 2-12% of patients with adenoma experience apoplexy
- Diagnosis of pituitary tumor unknown at time of apoplexy in ¾ of cases
- Presentation for 0.6-9.0% of surgically managed pituitary adenomas
- Male predominance ~60%
- Peak incidence in 5th decade


Pathophysiology
- Hemorrhage or infarction of a pituitary tumor causes a sudden enlargement of the gland due to ischemia and/or necrosis
- 2/3 are spontaneous
- 1/3 precipitating factor:
  - Hypotension
  - Surgery
  - Malignant hypertension
  - Anticoagulant treatment
  - Dopaminergic agonist treatment


Ocular Manifestations
- Visual Field Loss
  - Bitemporal
  - Junctional scotoma
  - Unilateral and/or bilateral ophthalmoplegia ~50%
  - CNIII>CNVI>CNIV

Symptoms evolve from hours to 2 days after onset of apoplexy

Systemic manifestations
- Sudden severe headache ~80%
  - Retroorbital
  - Bifrontal
  - Diffuse
  - Assoc with vomiting/nausea
  - Neck stiffness
- Brain stem/hypothalamus compression
  - Reduced consciousness
  - Thermoregulatory dysfunction
- Pituitary dysfunction
  - Thyrotropic deficiency
  - Hypothyroidism
  - Corticotropic deficiency
  - Hypercortisolism
- Hypothalamic dysfunction
  - Hypernatremia
- Hypotension
- Hypertension
- Hypercortisolism

Diagnosis
- CT scan ~46% sensitivity
- MRI – modality of choice

References

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