Cases of Sudden Vision Loss and Diplopia for the Primary Care Optometrist

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Neuro-ophthalmic Disease

- Relatively “new” compared to other subspecialties
  - Ancient Egyptians performed cataract surgery
  - Ancient Greeks developed salves for infections
  - Limited knowledge regarding eye movement and perception
  - 1820’s: believed globe moved around a fixed point at posterior surface
  - 1880’s: believed cerebral cortex had little, if any, role in perception
  - 1918: Gordon Holmes mapped retina on the visual cortex
  - 1927: Harry Moss Traquair describes our visual field as “an island of vision or hill of vision surrounded by a sea of blindness”
  - 1947: Dr. Frank Walsh published textbook on neuro-ophthalmology

Diplopia

- Diplous (Greek for double) + ops (Greek for eye)
- Often the first manifestation of many systemic disorders, especially muscular or neurologic disease
- Incidence in United States is unknown
- Eye hospital in London, UK reported the incidence of diplopia as the chief complaint in 1.4% of the presenting cases
- Polyplopia: the perception of more than 2 images
- Often a monocular phenomenon due to refractive aberrations


Sudden Vision Loss

- Incidence unknown due to variable presentations
- The most he most common cause of painless sudden visual loss is ischemia
- Other etiologies include infection, inflammation, trauma, compression
- Functional visual loss: decreased visual acuity or loss of visual field with no underlying physiologic or organic etiology
- Accounts for up to 5% of referrals to eye care providers
- 79% are female and 21% are male


Orbital Pseudotumor

- Second most common inflammatory process of orbit
  - Idiopathic orbital myositis
    - Female:male ratio 2.1
    - Mean age 47
    - 50% bilateral
  - Orbital pseudotumor
    - Female:male ratio 1:1
    - Mean age 45
    - 97% unilateral (except pediatrics)
  - Signs include diplopia, proptosis, conjunctival injection, chemosis, periorbital edema, ptosis
  - Cardinal sign is worsening pain with eye movement
  - Visual function usually normal in orbital myositis
  - Visual function may be decreased due to compressive optic neuropathy

- Pathogenesis unknown; however, likely immunologic
  - Orbital myositis has been associated with systemic lupus erythematosus, rheumatoid arthritis and Crohn's disease
  - May spontaneously resolve after 3 to 6 weeks
  - Treatment recommended to relieve pain, diplopia, limit muscle fibrosis and prevent recurrences
  - Dramatic improvement after 3 days of systemic steroids
Foster Kennedy Syndrome
- First described in 1911 by neurologist Robert Foster Kennedy
- Described compression of one optic nerve by a sub-frontal meningioma or olfactory groove tumor which results in compressive optic neuropathy (optic nerve head pallor) and increased intracranial pressure causing contralateral optic nerve head edema.

Pseudo-Foster Kennedy Syndrome
- Diagnosis of exclusion!
- Sequential anterior ischemic optic neuropathies most common
- R/O giant cell arteritis
- Check complete blood count with differential, erythrocyte sedimentation rate and C-reactive protein
- Modify vascular risk factors
- No other interventions have found to be effective

Subarachnoid Hemorrhage
- Hemorrhage between arachnoid membrane and pia mater
- Accounts for up to 7% of all cerebrovascular accidents
- Symptoms:
  - "Thunderclap headache" – severe with rapid onset
  - Vomiting
  - Confusion
  - Altered mental status
  - Seizures
  - Up to 50% are fatal

Pituitary Adenoma
- Pituitary gland anterior lobe secretes 6 hormones:
  - thyroid-stimulating hormone (TSH)
  - adrenocorticotropic hormone (ACTH)
  - follicle-stimulating hormone (FSH)
  - leuteinizing hormone (LH)
  - growth hormone (GH)
  - prolactin (PRL)
- Posterior pituitary gland secretes vasopressin and oxytocin
- Most tumors arise from anterior lobe increasing in size creating both systemic & visual effects (due to impingement on anterior notch of chiasm)
- Tumor growth asymmetric, therefore visual field loss asymmetric

Suprasellar Craniopharyngioma
- Differentials: pituitary tumor, meningioma and aneurysm
- Develop from nests of odontogenic epithelium derived from Rathke's pouch
- Benign tumors, usually slow growing
- Two types:
  - Adamantinomatous craniopharyngiomas
  - Papillary craniopharyngiomas
### Optic Neuritis: Signs and Symptoms

- Acute vision loss (20/20 to NLP)
- Pain on eye movement (92.2%)
- Dyschromatopsia
- Uthoff's sign
- Afferent pupillary defect
- Visual field defect
- Mild optic nerve swelling
- Mild vitritis
- Pulfrich Phenomenon (altered depth perception)

### Typical Optic Neuritis

- 77% female
- Average age 32 years (18-46)
- Pain on eye movements in 92.2%
- MRI: Periventricular abnormalities in 59% vs. 35% when no clinical signs of multiple sclerosis
- Visual field defects:
  - 48% diffuse loss
  - 20% altitudinal, arcuate or nasal step
  - 8% central or centrocecal
  - 16% other

### Atypical Optic Neuritis

- Outside 18 to 46 year span
- Painless
- Bilateral
- Continues to worsen beyond 14 days of onset
- Presence of other causative condition
  - Connective tissue disease
  - Infections
  - Infiltrative
  - Evidence of retinitis, vitritis or uveitis

### Optic Neuritis: Differentials

- Viral infection
- Intra-ocular inflammation
- Malignant hypertension
- Diabetic papillitis
- Sarcoid
- Syphilis
- Tuberculosis
- Collagen Vascular Disease

### Treatment Options

- Acute episodes
  - High-dose intravenous methylprednisolone
- Immunomodulators for chronic therapy reduce number and severity of relapses
  - Interferon beta (IFNβ)
  - Glatiramer acetate (GA)
  - Natalizumab
- Gilenya™ (Fingolimod)
  - First FDA approved oral immunomodulating agent
  - Sequesters lymphocytes
  - High dosages have been associated with macular edema

### Optic Neuritis: Prognosis

- Good
- 70% recover to 20/20
- Typical optic neuritis:
  - 95% recover to 20/40 or better within 1 year
  - Complete recovery usually within 8 weeks
- Permanent subtle color deficits common
Optic Neuritis: Management

- No effective treatment of idiopathic optic neuritis
- If periventricular abnormalities on MRI
  - IV Methylprednisone 1 gram QD in divided doses for 72 hours
  - Then prednisone 1 mg/kg/day PO with taper over 11 days
    - Check glucose, PPD and chest x-ray first

Arachidonic Acid Pathway

Inhibited by corticosteroids

Important Considerations Before Prescribing

- Is the patient diabetic?
- Does the patient have any infectious disease such as syphilis or tuberculosis?
- Does the patient have any gastrointestinal problems such as peptic ulcers?
- Is the patient pregnant?

Oral Steroids

- Prednisone
  - Widely available and inexpensive
  - Dosage is individualized for the severity and nature of the condition.
  - Most ocular inflammatory conditions respond to an initial dose in the range of 40-60 mg and tapering over one to three weeks.
    - Example: 40 mg for two days, 30 mg for two days, 20 mg for two days, 10 mg for two days, 5 mg for two days.
- Methylprednisolone (Dosepak)
  - Composite of 4 mg tablets, six tablets (24 mg) are taken on day one and reduced by one tablet per day over the next six days.
  - 24 mg is suboptimum therapy in most instances where an oral steroid is indicated.
  - Dosepaks are expensive compared to generic prednisone.

Acute Disseminated Encephalomyelitis (ADEM)

- Inflammatory demyelinating condition
- Closely resembles multiple sclerosis (MS)
- Multiple sclerosis:
  - Usually a chronic relapsing and remitting disease of young adults
  - Abnormalities of findings on cerebrospinal fluid immunoglobulin studies
- ADEM:
  - Monophasic disease, often in pre-pubertal children.
  - Usually occurs after a febrile illness or immunization
  - Usually affects many systems of the body
  - Varies in degrees of encephalopathy
- Continuum with MS and others?

Conversion Disorder

- A mental health condition in which a person has blindness, paralysis, or other nervous system (neurologic) symptoms that cannot be explained by medical evaluation – National Library of Medicine
- Symptoms typically begin after stressful experience
- Patients are NOT malingering
- Requires psychological evaluation and therapy
Nasopharyngioma

- A squamous cell carcinoma, or undifferentiated type, most common in males
- Nasopharyngeal carcinoma is the most common cancer originating in the nasopharynx
- Multifactorial causes: viral, environmental influences, and heredity
- Staging
  - Stage I: small tumor confined to nasopharynx
  - Stage II: tumor extending in the local area, or that with any evidence of limited neck disease
  - Stage III: large tumor with or without neck disease, or a tumor with bilateral neck disease
  - Stage IV: large tumor involving intracranial or infratemporal regions, an extensive neck disease, and/or any distant metastasis

ESR and CRP

- In study looking at 119 patients with biopsy proven GCA:
  - ESR had a sensitivity of 86%
  - Elevated CRP had a sensitivity of 97.5%
  - Sensitivity of the ESR and CRP together was 99%
  - 1 of the 119 patients (0.8%) had normal ESR and normal CRP
  - 2 patients (1.7%) had elevated ESR and a normal CRP


Optic Neuropathy Work Up

- History
  - Rapid onset: optic neuritis, ischemic optic neuropathy, inflammatory, traumatic
  - Gradual onset (months): toxic / nutritional, compressive
  - Lengthy (years): compressive, hereditary
- Clinical Exam:
  - Visual acuity
  - Color vision
  - Visual field
  - Dilated fundus exam
- Auxiliary Testing:
  - Contrast sensitivity
  - Optical coherence tomography (OCT)
  - Electrophysiological tests
- Blood work
- Imaging

Orbital Fracture Management

Further treatment based on imaging results

- Medial Orbit: Observe and consider surgical repair if medial rectus entrapment
- Orbital Apex: Difficult to manage & controversial
- Orbital Roof: Neurosurgical consult
- Zygomatic: Observe vs. surgery
- Orbital Floor:
  - Systemic Antibiotics
  - Nasal Decongestants
  - Ice Compresses

Migraine Headaches

- Common cause of neuro-ophthalmic complaints
- Visual symptom most common
- Symptoms may also include photophobia, pupillary dilation and ophthalmoplegia

Sarcoidosis

- Abnormal collection of inflammatory cells (granulomas)
- 5-10% of those with sarcoidosis develop central nervous system involvement
  - Of those, 50-70% have abnormalities of cranial nerves
  - CN VII most commonly affected, has best prognosis
  - 1% of those will have neurosarcoidosis alone without involvement of any other organs
  - Seizures present in 15% of cases
  - 10% mortality
Tolosa-Hunt Syndrome

- Painful ophthalmoplegia caused by nonspecific inflammation of the cavernous sinus or superior orbital fissure
- Diagnosis of exclusion
- Corticosteroids are the treatment of choice, usually providing significant pain relief within 24-72 hours of therapy initiation

Thank You
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