Medical Implications of Sudden Monocular Blindness

Sudden Monocular Blindness

- Discuss the 8 causes of sudden blindness
- Salient symptoms and signs
- Focus on those entities with systemic implications especially for the internist/primary care physician
- Role of the Internist and Primary care physician in the evaluation and management of these disorders.

History

Temporal course

![Graph showing visual acuity over time with dates and diagnoses: Cancer, Glaucoma, Diabetes, AIDS, Tumor.](image)

Essential Bedside Eye Exam (for the non Ophthalmologist)

- PERLA and EOMI
- Visual Acuity
  - (best corrected or pinhole)
- Afferent Pupillary Defect
  - AKA: APD, Marcus Gunn pupil, swinging flashlight sign

Swinging Flashlight Sign

![Image of a flashlight being swung.](image)

Extended Bedside Eye Examination

- Inspect the eye / adnexae
- Fundus exam
  - Dilate with 2.5% mydriatic
  - discs and macula
- Eye movements
- Confrontation visual fields
Circulation of the Disc and Retina

- Central retinal a. occlusion
- Posterior ciliary a. occlusion
- CRA
- PCA
- Cilioretinal a. (25%)

Specialty Involvement in various causes of sudden monocular blindness

- Internist/Primary
- Ophthalmologist
- Neurologist

- Retinal artery occlusion
- Ischemic optic neuropathy
- Retinal vein occlusion
- Optic neuritis
- Vitreous Hemorrhage
- Retinal detachment
- Macular degeneration (Psychogenic)

Rhegmatogenous retinal detachment

- Ophthalmologist
- separation between sensory retina and RPE due to vitreous traction
- Peripheral retinal tears
- Trauma and myopia
- Associated light flashes floaters.
- sudden visual field loss
- Variable vision, +APD

Serous/Hemorrhagic PEDs

- CSR
- ARMD

Vitreous Hemorrhage

- Sudden, painless onset
- No APD
- Partial view or no view of the fundus.
- Due to
  - Neovascularization DM
  - Retinal Detachment
  - Trauma
  - SAH

Optic Neuritis

- Rapid vision loss over a period of hours to days
- 20-40 years
- Pain typically with eye movement
- Variable acuity
- APD
- VF loss
- Neurology: MRI, LP and steroids.
- Associated with MS
Retinal Artery Occlusions

- Sudden, painless onset
- + premonitory Amaurosis
- APD
- Fundus:
  - Milky white retinal edema
  - Cherry red spot
  - Gaps in blood columns
  - Normal disc
  - Complete (CRAO)
  - Sectoral (BRAO)
  - + emboli, vasculitis

Features on causes

- Frequently difficult to ascertain the precise mechanism based on the eye exam
- Most cases involve:
  - Local thrombosis due to atherosclerosis
- Less commonly:
  - Embolization
  - Vasculitis
  - Vasospasm
  - Hypoperfusion/hypotension

Associated conditions

- 90% systemic disease
- 65% hypertension
- 25% diabetes
- 25% cardiac valvular disease
  - More likely in patients <45
- 45% carotid atherosclerosis
  - 20% high grade stenosis

Retinal Artery Occlusion

Etiology

- Atherosclerosis, Carotid Disease
  - (Stenosis, Occlusion, Dissection)
- Cardiac
  - (Dysrhythmia, Valvular, SEE, Prosthetic Values, Mitral Myxoma, Cardiomyopathies)
- Embolus
  - (Calcific, Cholesterol, Platelet/Fibrin, Fat, Tumor, Septic, Air, FB)
- Vasculitis
  - (GCA, Lupus, Scleroderma, Churg-Strauss, PAN, Takayasu, Behcets,)
- Hypercoagulability/Blood DYSCRASIA
  - Inflammatory Bowel Disease, Essential Thrombocytosis, Leukemia, Protein C Deficiency, PLEURA, Oral CONTRACEPTIVES, HEMOCYTANIA, Anti-Phospholipid AB, HEMOCOAGULOPATHY,
- Miscellaneous
  - Carotid Cavernous Fistula, Migraine, Bruised, Ocular Hypertension, Posterior Arterial Loops.
- Trauma

Retinal Emboli

- Associated with increased mortality primarily from cardiac disease.
- 56% / 9 years compared to 27% in an aged matched controls
Multiple Branch Occlusions

Lupus

Antiphospholipid Ab Syndrome

BRAO with CNS findings

- 40 yo wm
- Multiple BRAO
- Mental status and other focal hemispheric signs
- Tinnitus

Microangiopathy of Brain and Retina (Susac's Syndrome)

Management of Retinal Occlusion

- Short term immediate treatment
- Urgent systemic workup
- Systemic treatment

CRAO: short term ocular treatment

- Emergent referral to an ophthalmologist
- Experimental occlusions: 90 minutes
- If the patient is seen within 8(?) hours of onset
  - Anterior chamber paracentesis
  - IV Diamox or Mannitol to lower IOP
  - 95% O₂ / 5% CO₂
  - Ocular massage to dislodge embolus
  - Anti fibrinolytic agents

CRAO: urgent systemic workup

- R/O diabetes, hypertension, hyperlipidemia, CAD
- Carotid evaluation:
  - Carotid duplex scan and/or MRA
  - Cerebral angiography for high grade stenosis.
- Cardiac evaluation
  - Cardiac echo
- Vasculitis:
  - ESR, ANA, Antiphospholipid antibody, temporal artery biopsy etc.
- Hematologic assessment especially in young patients

CRAO: systemic treatment

- Depends on the cause
- Consider the use of
  - Endarterectomy
  - Anticoagulation (Aspirin vs Heparin/Coumidan)
  - Valve surgery
  - Steroids, Immunosuppression
Anterior Ischemic Optic Neuropathy

- Occlusion of the posterior ciliary artery with optic disc infarction
- Optic disc is invariably swollen in the acute stage
- Retrobulbar ischemic optic neuropathy is rare. Diagnosis of exclusion after compression or infiltration are ruled out.

Fundus in AION

Non arteritic-AION
- 50-65 F or M
- PMH: hypertension (diabetes)
- painless, apoplectiform onset of monocular vision loss
- 20/20 to no light perception (NLP), Dyschromatopsia, APD
- Optic disc is invariably swollen in the acute stage
- Prognosis: slight improvement with persistent defects in vision
- second eye in 25 - 40% over 5 years
- There is no effective treatment
- Prednisone, ASA, Antiplatelets, Heparin and surgical fenestration have failed to show any benefits, ASA may reduce risk of second eye involvement.

Arteritic AION
- Most common cause of blindness in GCA
  - 95% AION 5% CRAO
- Adequate treatment must be started immediately to avoid second eye involvement.
- Occult GCA: normal ESR in 10 – 15% of patients with AION; sometimes without symptoms of PMR.

Etiology

- Nonarteritic AION
  - Hypertension
  - diabetes.
  - Anemia, blood loss,
  - Systemic hypertension
  - Malignant Hypertension
  - Renal failure
  - Radiation
  - Coagulopathy

- Arteritic AION
  - Giant cell arteritis (GCA)
  - other vasculitides

Other:
- Ocular: optic disc drusen, post op (cataract, glaucoma, LASIK)
- Misc: sleep apnea, glaucoma, migraine

Not: carotid or embolic!
## Comments on Case AN

- Second eye involvement in GCA
  - Within 1 week in >70% cases, untreated (or inadequately treated)
- What is adequate steroid coverage in AION/GCA at the start and during taper
- How urgently do you treat patients with GCA who complain of visual loss
- The need for communication between ophthalmologist and internist in the management of these cases
- Catastrophic implications for the patient and serious legal issues for the health care providers.

## Corticosteroids in the Treatment of GCA

- No studies have established the ideal dose of steroids
- No clear evidence that IV is more effective than PO corticosteroids (Hayreh et al. 2003)
- IV is indicated in patients with impending vision loss (premonitory amaurosis fugax, unilateral vision loss with or without early signs in the contralateral eye)
- Anecdotal reports of reversal of vision loss on IV Solumedrol
- Oral prednisone 80-100mg with vision loss (at least).
- Solumedrol 1 gm IV PB QD for 3-5 days (∼ 2 gms and 4 gms have been given) followed by po pred or...
- Dexamethasone 150mg q8 x 3-5 days followed by po pred

## Corticosteroids in the Treatment of GCA

### How long to treat

- There are no hard rules.
- Based primarily on ESR and CRP. Symptoms are used but not always reliable indicator of visual complications.
- Maintain high dose of po prednisone until ESR and CRP reach its lowest stable value (usually 2 weeks); then start gradual taper (10mg/month).
- Frequent followup intervals in the first 3 months or down to low stable maintenance dose.
- Maintenance dose (5 mg – 7.5mg) for 1-2+ years.
- If steroids fail, consider Azothioprine, MTX, Cytoxan or cyclosporin.


- Vision can deteriorate in 5-15% of patients on steroids
- Deterioration while on adequate doses of steroids usually develops within the first 5 days.
- Thrombocytosis may be a risk factor for progression
- Many examples in the literature of second eye progression despite the use of prednisone and IV Solumedrol
Middle aged woman with sudden blindness

- 44 yo wf; no medical problems
- h/o uncomplicated liposuction of thigh, belly and flank under general anesthesia
- "usual post op ecchymosis"
- 48 h later noted sudden, painless field loss od
- 20/20 ou, apd od, inferior altitudinal field losses od
- Hct 18, Hgb 6

Elderly man with sudden blindness

- 64 year old wm, sudden painless, vision loss OD
- 20 pound weight loss/6 months on a diet, fatigue, no headaches
- Exam
  - 20/40 OD, 20/25 OS, APD OD
  - Altitudinal visual field loss OU
- Blood pressure 150/80 mm Hg.
- Hct 15; Hb 4.5
- Chronic Renal Failure.

Pseudo Foster Kennedy Syndrome in 16 yo Male

Anti Phospholipid Antibody Syndrome

Sudden sequential vision loss and headaches in 19 yo Male

Acute Hypertensive Neuroretinopathy

BP 220/160, Pheochromocytoma

Medications implicated in AION

- Medications for Erectile dysfunction
  - Viagra, Cialis, Levitra
- Amiodarone
- Interferon beta

Erectile Dysfunction Drugs

- 43 reported cases: 38 viagra; 4 Cialis; 1 Levitra
  - 1 case with rechallenge history
- 170 million prescriptions taken by 23 million men
- 100 clinical studies; n=13,000 patients, no AION; Most patients had other risk factors for AION

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<thead>
<tr>
<th>Conclusion: “Probable” (not “certain”)</th>
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<td>Contributory factor in a multifactorial disorder</td>
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<th>FDA recommendations:</th>
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<td>Stop taking med with sudden vision loss</td>
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<td>Discuss potential increased risk with patients prior AION (P&gt;A)</td>
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<tr>
<td>Avoid in patients with prior unilateral AION or significant retinovascular disease</td>
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Amiodarone-associated Optic Neuropathy

- α β antagonist for cardiac arrhythmias
- ? ~1.79% of patients on the medication
- Insidious bilateral disc edema, normal vision, big blind spots
- "possible" link (not "probable" nor "certain")
- Benefits far outweigh the risk
- Cardiologist should decide based on risks of discontinuation, alternatives

Interferon alpha – associated Optic Neuropathy

- Antiviral, antitumoral, antiangiogenic, immunomodulatory
- Hepatitis C, Leukemias, Myeloma, Thrombocytosis
- Reversible, asymptomatic, dose-related vascular retinopathy
- Anecdotal evidence for AION
  - 12 cases, 7 bilateral
  - 1w – 7 months after starting the drug
- Possible association.

Important points for the internist

- Do not lower pressure too aggressively
- Cautious steroid taper
- Insist on disc edema, otherwise consider other causes like tumor.
- Workup those cases with atypical features:
  - Young
  - Bilateral
  - Constitutional symptoms/systemic disease

Retinal Vein Occlusions

- Elderly
- Painless, sudden loss
- Variable acuity, ± APD
- Distinctive, if not pathognomonic fundus findings.
- Unilateral

Retinal Vein Occlusion

- Glaucoma (25-70%)
- HBP(35-50%)
- Diabetes(10-15%)
- Hyperlipidemia (10%)

Most patients have no other systemic disorders; however young patients, bilaterality, thrombotic history or the presence of phlebitis should lead to a more extensive evaluation.

- Blood dyscrasias:
  - multiple myeloma, Waldenstrom, Leukemia, P Vera, Thrombocytosis, cryoglobulinemia, sickle cell
- Coagulopathy:
  - Antiphospholipid antibody, Protein C and S deficiency, APC resistance, anionex, pregnancy
- Retinal vasculitis (periphlebitis):
  - sarcoidosis, Eales disease, Behcet’s, uveitis
- Other:
  - Carotid cavernous fistula, retrobulbar anesthesia.
CRVO with Blood Dyscrasias

26 yo WM

Summary:
- sudden monocular blindness

- Retinal artery occlusions:
  - Branch, Central

- Ischemic optic neuropathy
  - Nonarteritic, Arteritic

- Retinal vein occlusions
  - Branch, Central

- Etiology/workup
  - Carotid / cardiac
  - HBP, DM
  - Vasculitis
  - Hypercoagulability
  - Blood dyscrasias

- Optic neuritis (pain)
- Ocular: vitreous hemorrhage, ARMD, Retinal detachment, psychogenic.