Medical Implications of Sudden Monocular Blindness

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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

Visual Acuity
20/20

Cataracts, Glaucoma,
Diabetes, ARMD, Tumor
Optic neuritis
Vitrous hemorrhage
Usual
AION, CRAO, CRVO
ARMD: Retinal Detachment

20/200

min-hrs days weeks months

Essential Bedside Eye Exam
(for the non Ophthalmologist)

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

Swinging Flashlight Sign

Extended Bedside Eye Examination

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

- Central retinal artery occlusion
- Posterior ciliary artery occlusion (25%)

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

Vitreous Hemorrhage

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

Specialty Involvement in various causes of sudden monocular blindness

<table>
<thead>
<tr>
<th>Internist/Primary</th>
<th>Ophthalmologist</th>
<th>Neurologist</th>
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</thead>
<tbody>
<tr>
<td>Retinal artery occlusion</td>
<td>Ischemic optic neuropathy</td>
<td>Optic neuritis</td>
</tr>
<tr>
<td>Retinal vein occlusion</td>
<td>Vitreous Hemorrhage</td>
<td>Retinal detachment</td>
</tr>
<tr>
<td>Macular degeneration</td>
<td>Macular degeneration</td>
<td>Macular degeneration (Psychogenic)</td>
</tr>
</tbody>
</table>

Serous/Hemorrhagic PEDs

- CSR
- ARMD

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

- ATHEROCLEROSIS, CAROTID DISEASE
  - (stenosis, occlusion, dissection)
- CARDIAC
  - (tachycardia, valvular, see prosthetic valves, Mi, myxoma, cardiomyopathy)
- EMBOLUS
  - (calcific, cholesterol, platelet/fibrin, fat, tumor, septic, air, FB)
- VASCULITIS
  - (GCA, lupus, ITP, churg-straus, PAN, TAKAYASU, Behcets,)
- HYPERCOAGULABILITY/BLOOD DYSCRASIA
  - (inflammatory bowel disease, essential thrombocythemia, leukemia, protein C deficiency, P VERSA, oral contraceptives, homocystinemia, anti phospholipid AB, hemolymphohatopathy)
- MISCELLANEOUS
  - CAROTID Cavernous fistula, migrane, drusen, ocular hypertension, prepatial arterial loops
- TRAUMA

Retinal Emboli

- 96% carotid
- 4% cardiac
- Retinal arterial emboli
  - associated with increased mortality primarily from cardiac disease.
  - 56% / 5 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 a 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% O\textsubscript{2} / 5% CO\textsubscript{2}
  - ? 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

Anterior Ischemic Optic Neuropathy

Etiology

• Nonarteritic AION
  - Hypertension
  - Diabetes.
  - Anemia, blood loss,
  - Systemic hypotension
  - 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!

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.
GCA: AN 167152

<table>
<thead>
<tr>
<th>WK</th>
<th>Clinical History</th>
<th>Vision</th>
<th>Steroids</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>72 WF, Headaches, ESR = 105, positive biopsy for GCA, Headaches resolved with treatment.</td>
<td>Normal</td>
<td>80mg</td>
</tr>
<tr>
<td>5w</td>
<td>Steroids gradually tapered</td>
<td>NLP OD</td>
<td>30 mg</td>
</tr>
<tr>
<td>+3d</td>
<td>New onset scotoma OD (ophthalmologist) Calls to report deterioration of vision OD. Told by ophthal to come in on Monday. Continue prednisone</td>
<td>NLP OD</td>
<td>30 mg</td>
</tr>
<tr>
<td>6w</td>
<td>Phone call. Vision in the unaffected eye OS is transiently blacking out. “my vision is worse” Told by the ophthal to come in tomorrow</td>
<td>NLP OD</td>
<td>1gIV</td>
</tr>
</tbody>
</table>

Admitted for IV solumedrol

AN 167152

<table>
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<tr>
<th>+3 m</th>
<th>OD</th>
<th>OS</th>
</tr>
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<tbody>
<tr>
<td>Vision</td>
<td>NLP</td>
<td>NLP</td>
</tr>
<tr>
<td>Tension</td>
<td>10</td>
<td>12</td>
</tr>
<tr>
<td>Pupils</td>
<td>Sluggish</td>
<td>Sluggish</td>
</tr>
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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 (7-2 gms and 4 gms have been given) followed by po pred.
- Dexamethasone 150mg q8 x 3-5 days followed by po pred


- 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

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 the 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 Azathioprine, MTX, Cytoxan or cyclosporin
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.

PseudoFoster 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
  - Conclusion: “Probable” (not “certain”)  
    - Contributory factor in a multifactorial disorder
- FDA recommendations:
  - Stop taking med with sudden vision loss
  - Discuss potential increased risk with patients prior AION (? A5)
  - Avoid in patients with prior unilateral AION or significant retinovascular disease
Amiodarone-associated Optic Neuropathy

- $\alpha \beta$ 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

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

Retinal Vein Occlusion

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

Vein Occlusion Associations

- 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, scle cell
- Coagulopathy:
  - Antiphospholipid antibody, Protein C and S deficiency, APC resistance, estrogen, pregnancy
- Retinal vasculitis (periphlebitis):
  - sarcoidosis, Eales disease, Behcet’s, uveitis
- Other:
  - Carotid cavernous fistula, retrobulbar anesthesia.
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.