Differential Diagnosis Of The Swollen Optic Disk

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Disclosures – Dr Eric Schmidt

- Allergan – Consultant/Speaker
- Aerie – Consultant/Speaker
- Kala Pharmaceuticals- Consultant
- Sun Pharmaceuticals- Speaker/ Consultant
- Zeiss- Speaker
- Eyenovia – Consultant
- Oyster point - Consultant

Correct diagnosis of swollen optic disk is critical:

- Can save vision in 1 or both eyes
- Can save the patient's life
- Underlying etiology must be discovered

- History and appearance lead you to correct diagnosis

History

- Age
- Severity of vision loss
- Systemic problems
- Medication
  - Steroids
  - Vitamin A
  - Hormones
  - Tetracycline
  - Nalidixic acid
  - Isotretenoin
- Race
- Patient description
- Pain
- Associated symptoms

Examination

- VA
- Pupils
- EOM
- Ancillary tests
- Slit Lamp
- Fundus
- IOP
- Optic nerve head exam!!!!

Optic Nerve Head Evaluation

- Disc margin
- Neuroretinal rim
- Nerve Fiber Layer
- Cupping
- Color
- Vessels
  - Dilated/attenuated
  - SVP
  - Shunt/Neovascular

Causes of swollen optic nerves

- Elevated ICP
- Vascular/ Ischemic
- Inflammatory
- Systemic
- Orbital tumors
- Brain Tumors
- Ocular Disease

Papilledema

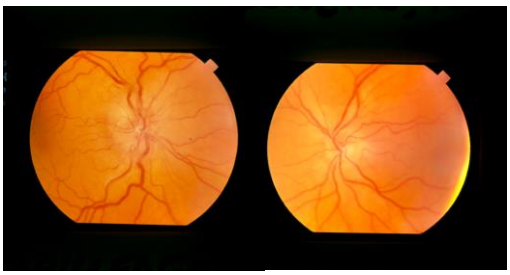
- Bilateral Optic Disk swelling due to increased intracranial pressure
  - Intracranial masses
  - Meningitis
  - Pseudotumor cerebri

Papilledema pathophysiology

- Flow of axoplasm is interrupted
- Axoplasm then accumulates at the lamina cribrosa
- Appearance of swollen optic nerve head ensues

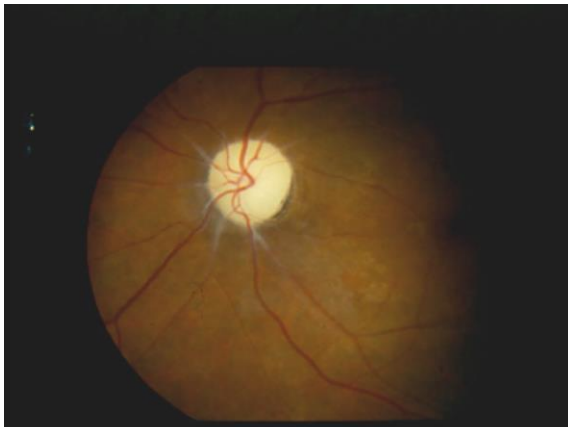
Papilledema

- Bilateral, may be asymmetrical
- NFL opacification
- Disc hyperemia
- SVP absent
- Cup preserved
- Splinter hemes
- CWS and exudates
- Hazy retinal vessels
- Paton's lines
- Appearance varies over time



Papilledema

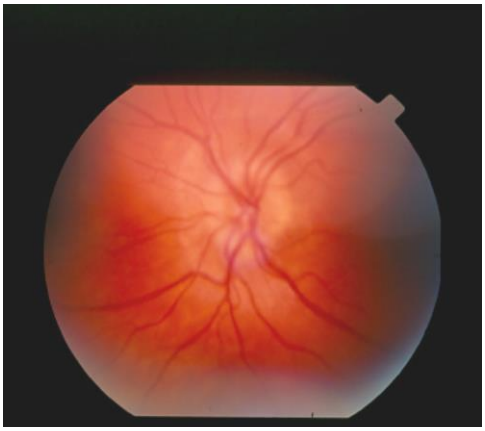
- VA is usually normal, however:
  - Recurrent TVOs may occur
  - TVO precipitated by postural change
  - VA may become very bad
- VF abnormal:
  - Enlarged blind spot
  - Generalized decreased mean sensitivity
  - May become "classic"
- Headaches are common



Papilledema is a true medical emergency!!!

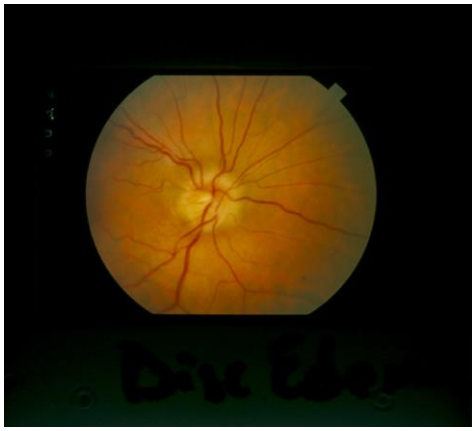
- CT scan/MRI to r/o mass lesion
- If normal then lumbar puncture

- Treatment depends upon cause
- Who should control the course?



Idiopathic intracranial hypertension – (BIIH, PTC)

- Papilledema plus-
  - HA, diplopia, and/or TVO
  - Increased CSF (otherwise normal)
  - Normal CT scan
  - Neurologically intact
  - "Classic" patient



IIH- Causes

- Obesity
- Dysmennorrhea
- Mediastinal mass
- Toxemia
- Idiopathic
- Viral meningitis
- Medications
- Sarcoid/SLE
- Syphilis
- Get blood work!!

Pseudotumor cerebri: Not-so-benign-intracranial hypertension

- Treatment
  - Acetazolamide 500mg BID
  - Prednisone 10-40mg QD
  - Repeat LP
  - Shunt surgery
  - Topamax
- Treatment
  - Weight loss
  - WEIGHT LOSS
  - **WEIGHT LOSS**
  - **WEIGHT LOSS!!!**
  - Lose weight

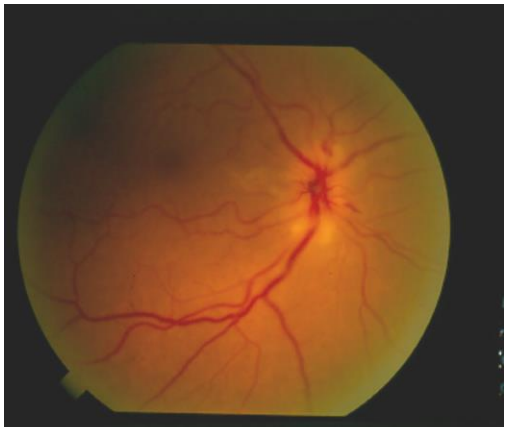
- Bilateral disc edema should be considered papilledema until proven otherwise
  - Get CT scan, if normal LP

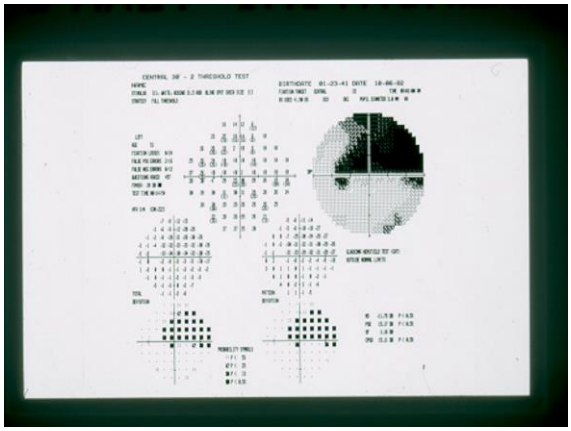
Ischemic Optic Neuropathy

- Acute, profound, painless loss of vision in older patients
- Affects one eye…*first!*
- May have associated symptoms
- Etiology – infarct of ciliary circulation within the laminar region

ION Appearance

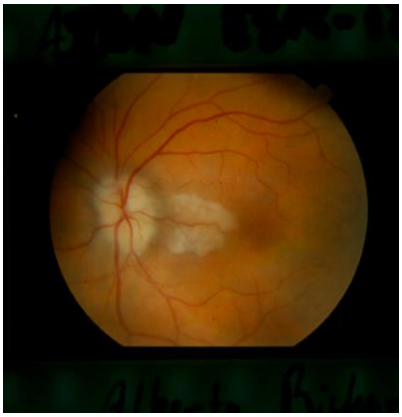
- Profound VA loss
- (+) APD
- Altitudinal hemianopsia
- Splinter disc hemes
- Cotton wool spots
- Pallid disc edema
- Vision does not improve
- ***Must differentiate between arteritic and non-arteritic***

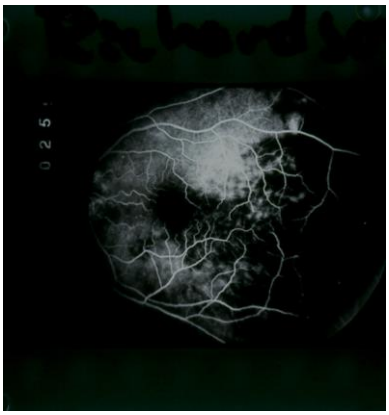




Arteritic ION – (GCA)

- 70-80 y/o
- Severe VA loss
- 2nd eye involvement 75%
- Disk is less hemorrhagic
- OIS, CRVO, CRAO
- Systemic – HA, PMR, jaw claudication
- ESR elevated
- Marked steroid response





Giant Cell Arteritis

- Autoimmune disorder affecting blood vessels
- Pxs over 60 y/o
- Severe, pulsating HA
- Scalp tenderness
- Jaw claudication
- PMR
- Malaise
- Low grade fever
- ELEVATED ESR/CRP

GCA

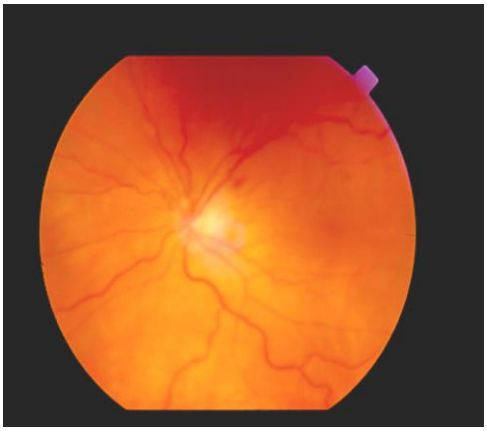
- 2nd eye involvement
- May have a prodrome
- Tender, palpable, non-pulsatile arteryu
- Markedly elevated ESR
- TRUE OCULAR EMERGENCY!!

ION should be considered a true ocular emergency

- Think GCA until ruled out
- STAT ESR/CRP
- Associated symptoms
- Temporal artery biopsy
- Prompt oral steroids can save other eye

Non-arteritic ION

- 60-70 y/o
- More moderate VA loss
- 2nd eye 40%
- Appearance – more hemes and disk edema
- Normal ESR
- Poor steroid response
- Etiology – hypertension/embolic
- Idiopathic?
- Tx – Blood thinners
- Statins
- CV referral



Ischemic optic neuropathy

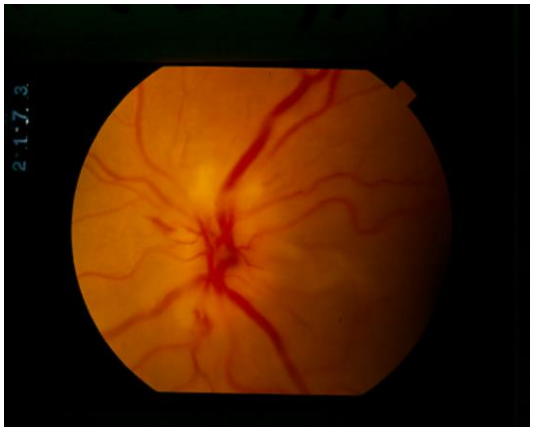
- Arteritic
  - 70- 80
  - Severe VA loss
  - 2nd eye 75%
  - PMR
  - Steroids
  - Elevated ESR
- Non-arteritic
  - 60 -70
  - Moderate VA loss
  - 2nd eye 40%
  - Hypertension
  - Decrease BP
  - Normal ESR

Optic Neuritis

- Primary inflammation of the optic nerve
  - 20 -50 y/o
  - Unilateral
  - (+) APD
  - Pain on eye movement
  - Rapid loss of VA over 7 day period, may improve over time

Optic neuritis appearance:

- Many hemorrhages and hard exudates
- Cupping obliterated
- Vitreous cells
- May be sectoral
- Classic altitudinal hemianopsia
- Residual appearance may show pallor



VF in Optic Neuritis

- Central field affected 97%
- Peripheral fields affected 70%
- Peripheral field recovers more fully and more completely

Optic Neuritis Etiologies

- Idiopathic
- MS
- Measles, mumps
- Granulomatous inflammation
- Viral infection (HZ, mono)
- Contiguous inflammation of sinus or meninges
- Intraocular inflammation

Retrobulbar Optic Neuritis

- Disease in which neither the patient nor the doctor sees anything!
- (+) APD
- Profound vision loss
- Otherwise normal exam
- Infarction of retrolaminar circulation

Optic Neuritis Therapeutic Trial (ONTT)

- Acute cases of ON, if VA 20/40 or better:
  - Observe
- If VA 20/50 or worse:
  - IV methylpred 250mg given over 30 min Q6H for 12 doses followed immediately by:
  - Oral pred 1mg/kg/day for 11 days then
  - 20mg pred for 1 day then,
  - 15mg pred QOD

ONTT – 30 Years Later

- Oral Steroids are not used for isolated ON
- MRI established as most important risk factor for development of MS
- Low risk profile for MS identified
  - Normal MRI with poor vision (esp men)
  - No pain
  - Severe disk swelling
- VF established as essential
- Fellow eye is at risk
- IV corticosteroids early on save vision (1 line at 3 weeks)
- Inverse relationship confirmed between VA loss and neurological deficits

Multiple Sclerosis

- Most common neurologic disease
- Affects young white females 3:2
- Geographic preponderance
- Marked by variable course, remissions and exacerbations
- Demyelinating disease with unknown cause

MS – most frequent presenting symptoms

- Motor weakness
- Paresthesia
- Visual/ocular complaints (40)%
- ataxia

MS Ocular Signs

- ON or RON *******
- Bell's palsy
- Internuclear ophthalmoplegia
- Cranial nerve palsy
- Uthoff's sign –pathognomonic for MS
- Visual field defects

MS- initial ocular symptoms

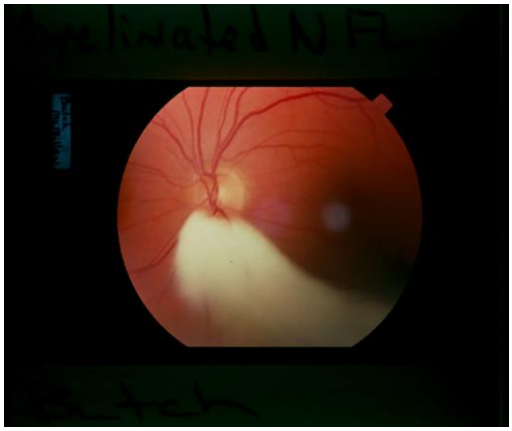
- Blurred vision – 47%
- Lagophthalmos- 20%
- Diplopia – 17%
- Eye pain – 17%
- Nystagmus – 4%
- Dyschromatopsia
- Color desaturation
- Decreased contrast sensitivity

In a younger person with optic neuritis:

- Suspect MS
- Get good history
- Check for associated neurological defects
- Follow closely
- Get MRI
- To tell or not to tell

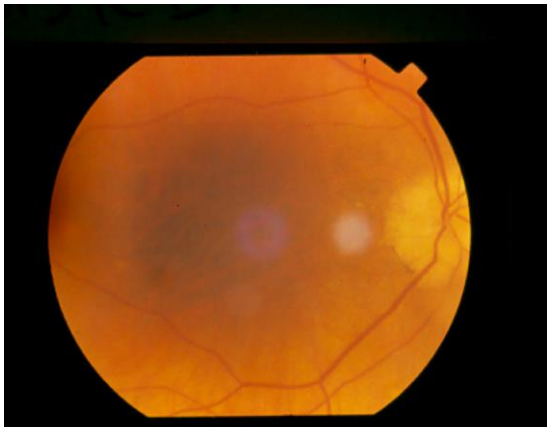
Pseudopapilledema

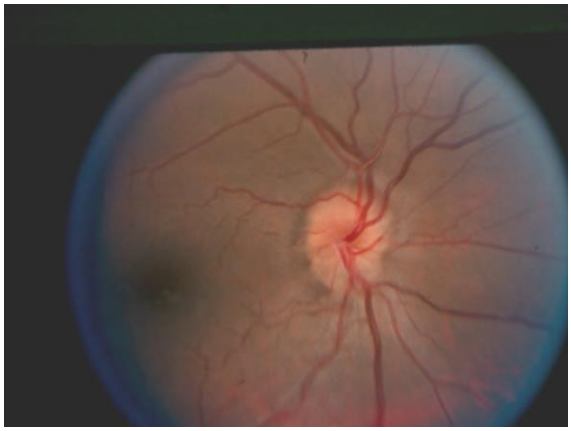
- Myelinated NFL
- Hyperopic disk
- Diabetic papillopathy
- Tilted disk
- Hypertensive papillopathy
- Optic disk drusen
- CRVO



Optic disk drusen

- Buried hyaline bodies
- Scalloped borders
- Bilateral
- Asymmetrical elevation
- Absence of cup
- Rare hemorrhages
- B-scan
- Can get VF loss, SRNVM
- VA may be affected





Compressive optic neuropathy

- Optic disk swelling and <u>decreased VA</u>
  - Caused by compression of optic nerve
  - Grave's disease
  - Optic nerve tumors
  - Orbital inflammation
  - Vascular inflammation
  - Trauma
- Watch pupils, proptosis and color vision

Papillophlebitis

- Swollen optic nerve head in younger patient
- Fairly normal VA
- Subjective "blur"
- No APD
- Enlarged blind spot
- Self-limiting
- Type "A" personality ?