

Differential Diagnosis Of The Swollen Optic Disk

Eric E. Schmidt, O.D., F.A.A.O.
Omni Eye Specialists
Wilmington, NC
schmidtvision@msn.com

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 | ▪ Race |
| ▪ Severity of vision loss | ▪ Patient description |
| ▪ Systemic problems | ▪ Pain |
| ▪ Medication | ▪ Associated symptoms |
| ▪ Steroids | |
| ▪ Vitamin A | |
| ▪ Hormones | |
| ▪ Tetracycline | |
| ▪ Nalidixic acid | |
| ▪ Isotretinoin | |

Examination

- | | |
|-------------------|-----------------------------|
| ▪ VA | ▪ Slit Lamp |
| ▪ Pupils | ▪ Fundus |
| ▪ EOM | ▪ IOP |
| ▪ Ancillary tests | ▪ 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

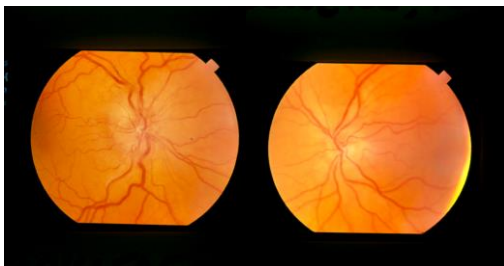
- Bilateral Optic Disk swelling due to increased intracranial pressure
 - Intracranial masses
 - Meningitis
 - Pseudotumor cerebri

Papilledema pathophysiology

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

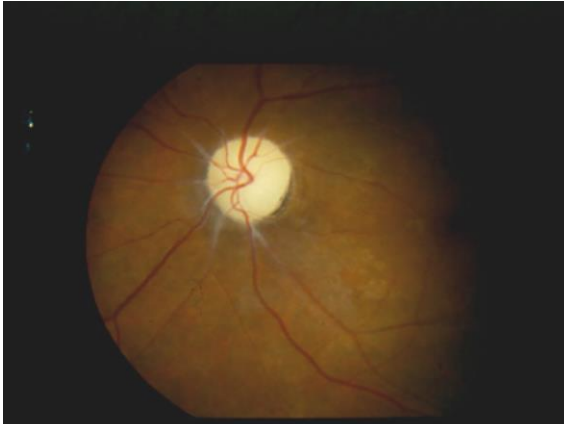
Papilledema

- 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

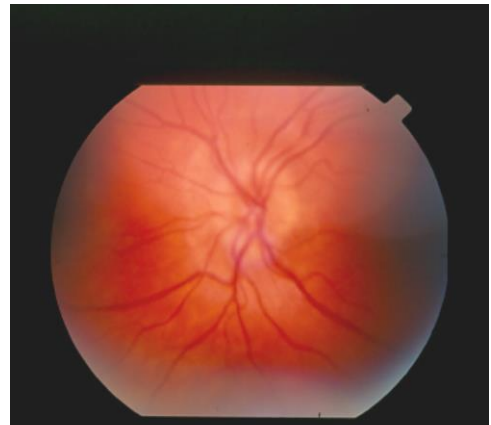
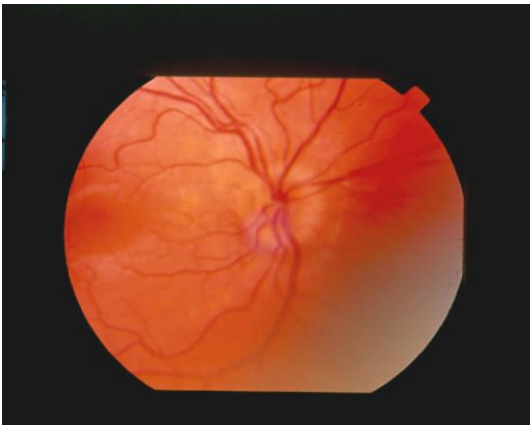
- VA is usually normal, however:
 - Recurrent TVOs may occur
 - TVO precipitated by postural change
 - VA may become very bad
- VF abnormal:
 - Enlarged blind spot
 - Generalized decreased mean sensitivity
 - May become “classic”
- Headaches are common



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

- | | |
|--|---|
| <ul style="list-style-type: none"> ▪ Treatment <ul style="list-style-type: none"> ▪ Acetazolamide 500mg BID ▪ Prednisone 10-40mg QD ▪ Repeat LP ▪ Shunt surgery ▪ Topamax | <ul style="list-style-type: none"> ▪ Treatment <ul style="list-style-type: none"> ▪ Weight loss ▪ WEIGHT LOSS ▪ WEIGHT LOSS ▪ WEIGHT LOSS!!! ▪ Lose weight |
|--|---|

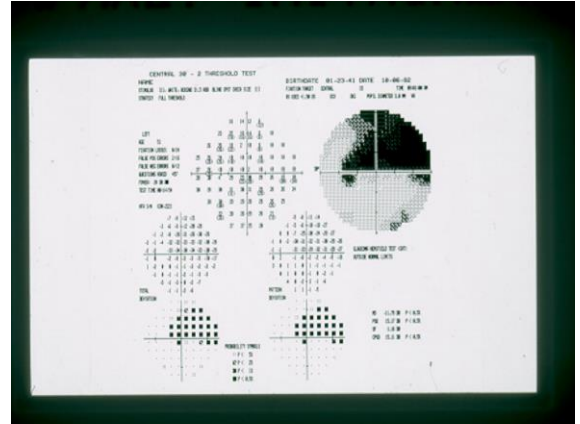
- Bilateral disc edema should be considered papilledema until proven otherwise
 - Get CT scan, if normal LP

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

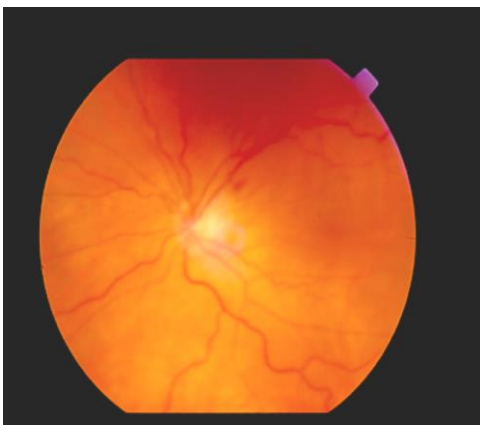
- 2nd eye involvement
- May have a prodrome
- Tender, palpable, non-pulsatile artery
- Markedly elevated ESR
- TRUE OCULAR EMERGENCY!!

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

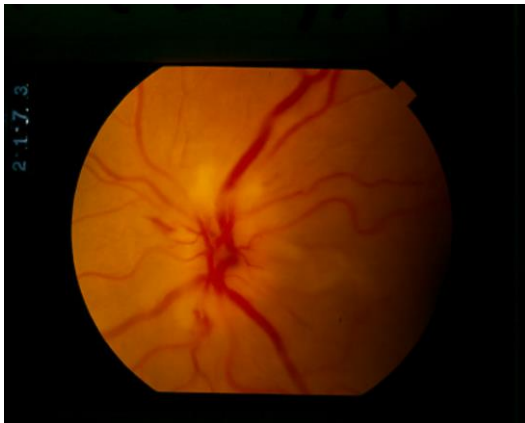
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| <ul style="list-style-type: none"> ▪ Arteritic <ul style="list-style-type: none"> ▪ 70- 80 ▪ Severe VA loss ▪ 2nd eye 75% ▪ PMR ▪ Steroids ▪ Elevated ESR | <ul style="list-style-type: none"> ▪ Non-arteritic <ul style="list-style-type: none"> ▪ 60 -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

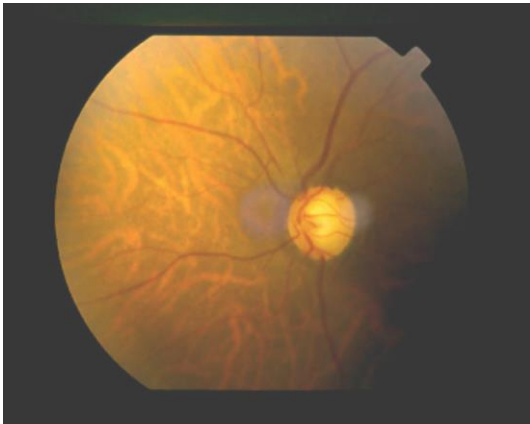
- Central field affected 97%
- Peripheral fields affected 70%
- Peripheral field recovers more fully and more completely

Optic Neuritis Etiologies

- | | |
|--|--|
| <ul style="list-style-type: none"> ▪ Idiopathic ▪ MS ▪ Measles, mumps ▪ Granulomatous inflammation | <ul style="list-style-type: none"> ▪ 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

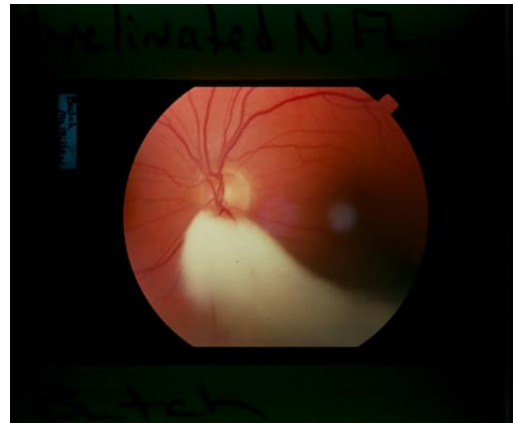
- Blurred vision – 47%
- Lagophthalmos- 20%
- Diplopia – 17%
- Eye pain – 17%
- Nystagmus – 4%
- Dyschromatopsia
- Color desaturation
- Decreased contrast sensitivity

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

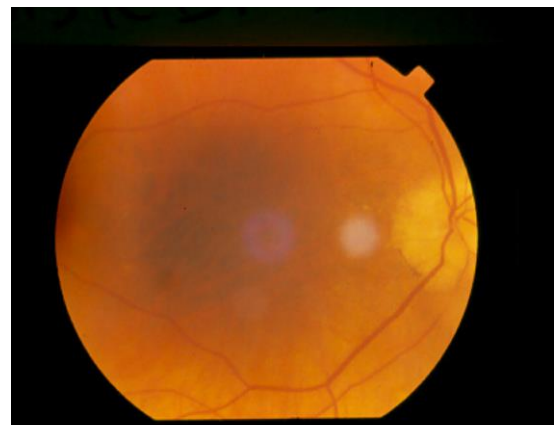
Pseudopapilledema

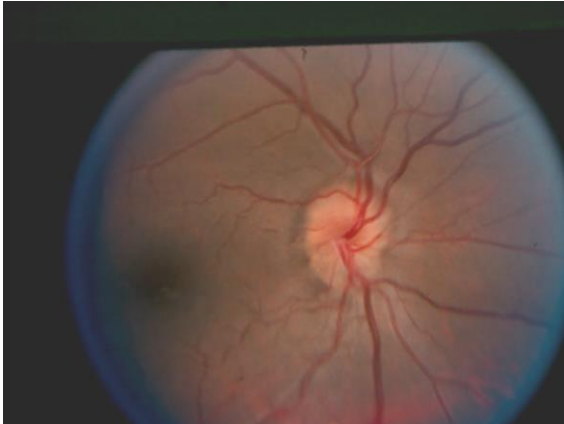
- Myelinated NFL
- Hyperopic disk
- Diabetic papillopathy
- Tilted disk
- Hypertensive papillopathy
- Optic disk drusen
- CRVO



Optic disk drusen

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





Compressive optic neuropathy

- Optic disk swelling and decreased VA
 - 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 ?