Don’t Let Swollen Optic Nerves Make You Nervous

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Financial disclosures

- No financial disclosures
Examination Techniques

- Stereoscopic viewing essential
- VA and VF: SVP/EVP
- Pupil testing and color vision
- Brightness comparison and red cap test
Papilledema

- Bilateral* optic nerve head swelling secondary to increased ICP
- Swollen, blurred margins with splinter hemorrhages and exudates as well as nerve fiber layer edema. Patton’s folds may be seen: chorioretinal folds concentric around the disc
Papilledema

- May be asymmetric or very rarely unilateral (sequential swelling—one nerve can swell a few weeks or months before the other)
- VA varies but typically mild reduction only or no loss at all
- May get diplopia secondary to abducens nerve compression
- With increased ICP, can get choroidal folds only (before papilledema) at lower pressure levels
Papilledema

- VF usually shows enlarged blind spot
- No pupillary defect. Normal color vision
- SVP absent with obliterated cup
Papilledema from idiopathic intracranial hypertension (IIH)
Papilledema IIH age 15
OCT
Papilledema (HTN)
Papilledema (tumor)
Subtle papilledema (IIH)
Papilledema IIH

Patton’s folds
Papilledema IIH
Terson’s (bleeding from sub-arachnoid hemorrhage) and papilledema
Papilledema progression
Patton’s Folds
Patton’s folds: now you see them in 2014……
Back then in 2007 you did not...
Patton’s folds
Longstanding papilledema with optic atrophy (IIH)
Papilledema OCT NFL

The image shows OCT scans and tables with measurements related to papilledema. The scans indicate increased pressure in the optic nerve head. The tables list various measurements such as mean, SD, and range for different parameters.
NFL edema
Increased ICP

- Variations are due to anatomical considerations
- If the channels connecting the central cavity and optic nerve sheath allow equal flow on both sides and in both directions, papilledema will occur and will improve with decreased ICP
Increased ICP

- If there is a difference in the communications then the edema will be asymmetric. Usually the result of a smaller bony canal opening on one side limiting the swelling.
- If the valves are one-way then the swelling will not improve rapidly with Tx
Increased ICP

- An acute rise in ICP that resolves rapidly is not typically associated with papilledema. Elevation must be chronic.
- Increased pressure is transmitted from the sub-arachnoid space to the optic nerve head via the nerve sheath. Venous pressure in CRV increases.
- Disruption in axoplasmic flow at lamina cribosa leads to swelling.
Etiologies of Increased ICP

- Space occupying lesion; must always be ruled out!
- Infection or anatomical abnormality
- Malignant hypertension
- IIH
- Certain medications (tetracycline, high dose vitamin A, steroids, Accutane, others)
- ? Sleep apnea (obesity): ICP may be elevated only at night! Men especially
- Must order MRI in all cases
Idiopathic Intracranial Hypertension (IIH)

- Older term is “pseudotumor cerebri”
- Young overweight females (F 8X M)
- 1 / 100,000 in population as a whole; 20 / 100,000 in 20 - 44 year old women 10% over ideal weight
- May be related to medications including TCN, HRT, lithium, high dose Vitamin A supplementation, steroid withdrawal
- Sleep apnea link
IIH

- Symptoms of transient blur, diplopia, tinnitus (intracranial noises, not just ringing), headaches, etc.
- ICP usually severely elevated; normal is 50 – 200 mmH20. Single measurement can be misleading: levels can vary over 24 hours.
IIH

- Diagnosis requires normal MRI / MRV and CSF studies with elevated ICP
- Watch for spinal chord tumors
- Watch for transverse venous sinus stenosis / thrombosis (MRV)
IIH Management

- Refer to a neurologist
- Medical management includes Diamox, Lasix, Topamax
- Weight loss
- NORDIC IIHTT: recruiting at 43 sites
IIH Management

- If recalcitrant....
- Repeated lumbar taps (ugh!)
- Lumbo-peritoneal shunt
- Ventricular shunt
IIH Management

- If progressive changes in visual acuity or visual field occur, consider an optic nerve sheath decompression.
- Several small fenestrations in the optic nerve sheath are created to allow room for expansion.
- Performed by a neuro-ophthalmologist. Often do worse eye only because 50% get improvement in the fellow eye.
Foster Kennedy Syndrome

- Swollen optic nerve on one side, advanced optic atrophy on the other
- Advanced optic atrophy prevents swelling making a bilateral problem appear to be unilateral
- Often seen in chiasmal tumors
Compressive Optic Neuropathy

- Compression leads to axoplasmic stasis and retrograde death of nerve fibers
- Pale, choked, swollen nerve
- Rarely see hemes; + APD
Compressive Optic Neuropathy

- Optic atrophy and severe vision loss with time
- MRI with and without contrast: neurosurgery referral
- Possibly endoscopic optic nerve decompression?
Pituitary tumor with compressive optic neuropathy OD
Pituitary tumor post surgery with neuropathy OU
Sphenoid wing meningioma with compressive neuropathy
ION

- Nonarteritic
- Arteritic
Nonarteritic ION

- Swollen, hyperemic nerve with splinter hemes and exudates
- Often sectoral
- Ischemic / hyoperfusion event caused by interruption of micro-vascular circulation, often at night.
- Highly associated with sleep apnea (75-90% in several studies)
- NAION has 5x risk of sleep apnea, 8x risk in women
NAION

- No systemic symptoms; normal ESR / CRP
- Most common cause of ONH swelling over the age of 55 (2-10 cases per 100,000 per year)
- 45-60 year olds (any age possible) with no sex predilection; C > AA
Nonarteritic Etiologies

- 1) Sleep apnea! Up to 90%
- 2) Hypertension (40%) (med related?)
- 3) Idiopathic (27%)
- 4) Diabetes (17%)
- 5) Atherosclerosis (14%)
- 6) Migraine (12%)
- 7) Increased Homocysteine / Decreased vitamin B6
- 8) HIV infection
Nonarteritic ION

“Disc at risk”: NAION typically found in nerves with very small / almost non-existent cups due to crowding and compression of axons.

Approximately 15% of cases will involve the fellow eye in 5 years (more common with VA < 20/200 in first eye, diabetics, and platelet polymorphisms). Repeat attacks in same eye < 5%.
NAION

- VA varies widely from normal to severe loss: 45% 20/40 or better but 33% 20/200 or worse
- VA loss progresses over 2-4 weeks
- VA improves by up to three lines at six months in 40%
- In patients under 50 years of age, there is a higher rate of bilateral involvement and more visual recovery
Nonarteritic ION

- Often APD, color vision usually normal
- Most frequent visual field defect is inferior nasal / partial altitudinal respecting the horizontal midline. FDT may be more sensitive and often shows spillover of loss in to “non-affected” hemifield
- After swelling resolves the nerve is pale but often not cupped-cupping may occur, however
- Why does area of swelling not always match VF defect? Because original swelling resolves quickly but causes secondary contiguous compressive swelling
NAION OD 2 weeks after onset of symptoms
Note how VF defect does not match area of swelling
Nonarteritic ION Treatment

- No treatment other than managing the underlying cause has proven to be consistently effective
- Blood thinners may debatably protect the fellow eye but will not alter the course of recovery.
- Order CBC, ESR and CRP, lipid profile, hemoglobin A1C. Check BP
- Check for sleep apnea in all cases!
Steroids?:

- SS Hayreh: 2008 study utilizing oral steroids....
- If VA 20/70 or worse, several week course of oral prednisone resulted in VA improvement (3 or more lines) in 70% of treated patients and 40% of untreated patients
- Beginning dose of 80mg for 2 weeks with slow taper.
Incipient ION

- We see it coming, but can we do anything about it? No, other than manage the underlying condition.
- Will it always end badly? No: sometimes it just resolves with no VA or VF loss.
NAION
NAION
NAION
NAION OD secondary to HIV
Old NAION OD
NAION OD: The Beginning

NAION

Fellow Eye
Optic atrophy / incipient ION

Optic Atrophy

Incipient NAION
Optic Atrophy

NAION OS
Optic atrophy OU

Post NAION

Post NAION
ION OS with matching VF / NFL loss

Spill over?
Arteritic ION

- Pale disc swelling with splinter hemorrhages
- Over 60 years old, F>M, Caucasians
- Increased ESR and C-Reactive protein
- ESR normal in about 25%!
- VA 20/200 or worse in 60% of cases
Arteritic ION

- Sudden, painless loss of vision with APD
- Altitudinal VF loss most common, others possible
- Symptoms of GCA but about 1/3 are symptom free
- Very high five year mortality rate
Giant Cell Arteritis

- GCA is a disease of unknown etiology (emerging evidence that zoster may be involved as trigger) affecting the large and medium arteries including the temporal, ophthalmic, and posterior ciliary arteries.
- Symptoms include HA, scalp tenderness, jaw claudication, malaise, fever, and fatigue.
GCA

- May also see CWS, CRAO, and amaurosis fugax
- 20% of cases with ocular involvement are CRAO, 80% ION
- Obtain stat Westergren ESR, CRP, CBC (anemia-false ESR)
Giant Cell Testing

- Normal ESR is age/2 for men and age +10/2 for women
- C-Reactive protein testing is not specific for GCA but it is nearly 100% sensitive so very useful test
- Temporal artery biopsy when indicated: can get false negatives due to “skip lesions”
Giant Cell Arteritis

- 25% of untreated patients develop AION
- 2/3 will develop in the second eye within weeks if not treated
- Rheumatology referral
Giant Cell Treatment

- IV hydrocortisone followed by long term oral prednisone. Maintenance dose of 10mg daily for years. Follow ESR, other markers
Amiodarone induced optic neuropathy

- Mimics NAION in nerve appearance but bilateral instead of unilateral
- Afflicts 2% of patients taking it
- Slow, insidious onset of visual loss
- Slow, complete recovery over many months after medication is discontinued (very long half-life)
- More common side effect of Amiodarone is vortex keratopathy
Viagra / Cialis / Levitra and NAION

- 553 cases officially reported to the FDA by the end of 2014. 443 were Viagra
- Under reported
- These medications also occasionally used for pulmonary HTN
- Visual loss most often noted upon awakening the morning after use
- Is the association real or coincidence?
- Likely the “straw that broke the camel’s back” in those with risk factors. But..................
ED drugs and NAION

- Very interestingly, has been reported in a 7 month-old infant, 28 year old, and 33 year old, presumably all taking them for pulmonary HTN
- At those young ages, not as likely to have other NAION risk factors
Viagra / Cialis

- What is the proposed mechanism? Nitrous oxide release actually dilates vessels.....but drops blood pressure.
- Do ION patients have faulty autoregulation?
- Ask all males with NAION about ED drug use. D/C if using to protect fellow eye.
Optic Neuritis

- Unilateral (usually) swollen nerve. Often retrobulbar (2/3) with no visible abnormality. Hemorrhages uncommon
- Diffuse visual field loss or enlarged blind spot. Subtle defects often present in the fellow eye
- Centro-cecal defect with Goldmann perimetry
- About 5% in US bilateral, but 30% in Asia
Optic Neuritis

- Younger patients (20-40 peak), F > M: more common in Caucasians
- APD, wide range of VA loss, decreased color vision; pain on eye movement (pain on eye movement not common in Asian patients)
Optic Neuritis

- Often associated with post viral syndromes or demyelinating diseases such as MS (initial MS symptom in 20% of cases-usually retrobulbar)
- VA recovers over weeks to months to near baseline level but often seems dim or washed out to the patient
- Get MRI in most cases
- May represent form fruste MS
- Several cases reported linked with use of TNF (tumor necrosis factor). Used for RA & JA: etanercept, infliximab, etc.
Optic neuritis and fellow eye
Optic neuritis with atrophy after six weeks
Optic neuritis associated with MS
Optic neuritis
Optic Neuritis Treatment Trial

- 457 patients in three treatment groups 1) oral steroids (1mg / kg / day X 14 days), 2) IV steroids (250mg Q 6h X 3 days) followed by orals (as above for 11 days), 3) placebo
- Orals followed by short taper of 20 mg on day 15 and 10 mg on days 16 and 18
- Hospitalized while on IV methylprednisone
- Traditional treatment of oral steroids proved to be the least effective of the three! Actually increased recurrence rate
ONTT

- IV followed by orals hastens VA recovery by about 2 weeks but does not improve end result.
- Delays the onset of MS symptoms up to 2-3 years: no benefit at 5 years.
ONTT 15-year F /U

- 294 patients seen 15 years out
- 15-year risk of developing MS was 50% (6% had known MS entering the trial)
- 72% if lesions on original MRI, 25% without
- VA 20/20 or better in 72%
- Factors indicating a lesser chance of developing MS include: 1) male gender, 2) optic disc swelling, 3) peripapillary hemorrhages and exudates, 4) no pain on eye movement, 5) NLP vision.............so many of these patients probably had something other than optic neuritis in the first place
Optic Nerve Head Drusen

- Increased prevalence in small nerves with small cups. Therefore, more common in whites than in AA. Higher incidence in patients with RP (10%)
- Compression of axons leads to stasis of axoplasmic flow and hyaline is excreted then calcifies over time, leading to the formation of drusen
- Nerve appears elevated but no splinter hemes or exudates and the margins are distinct.
- Abnormal vessel branching
Optic Nerve Head Drusen

- Not always visible! Buried early in life but become visible with time. Creation of more drusen push some forward to the surface of the nerve.
- Can cause decreased vision and variable visual field defects. More loss with visible drusen.
- Common and under diagnosed.
Optic Nerve Drusen

- SVP/EVP not affected: APD and color vision loss rare but possible
- Change with time
- Use B-scan or OCT to detect buried drusen
- Also seen with CAT scan, MRI, IVFA, and FAF (FAF better with visible drusen, limited for buried drusen)
B-scan and ONH drusen

- Gain must be turned down low to make drusen “stand out” against surrounding tissue
- Drusen show up as a hyper-reflective signal
- Not good in younger children because the hyper-reflective signal requires the drusen to calcify, and this has not happened yet at a young age
ONH Drusen
ONH Drusen
ONH Drusen
ONH Drusen
ONH Drusen
ONH drusen
ONH DRUSEN SD OCT
Color SD-OCT
FAF ONH Drusen
FAF ONH Drusen
NFL loss with ONH drusen: can cause VF defects
IIH with ONHD and true papilledema
IIH with ONHD and papilledema
Papilledema and ONH drusen

- Long standing papilledema can cause the formation of ONH drusen from axonal compression, leading to simultaneous involvement.
- Can be a real clinical challenge.
- Have to determine when active swelling is present and when it is not.
- NFL thickness scans on OCT are often very valuable.
ONH drusen MRI
ONH drusen B-scan
Papillophlebitis (optic disc vasculitis)

- An inflammatory variant of CRVO striking otherwise seemingly healthy, young adults (f 2x m)
- Disc edema out of proportion with retinal hemorrhaging
- Usually mild VA reduction to around the 20/30 level but can be worse
Papillophlebitis

- Vague prodrome of scintillating, colored lights with visual disturbances
- Enlarged blind spot on the visual field
- Dilated and tortuous veins
- Condition is self limiting over the course of several months and a complete recovery is the norm
- Can be associated with anti-phospholipid antibody syndrome, so consider blood work
Papillophlebitis
Diabetic Papillitis

- More common in young, type I diabetics but can also be seen in adults with type II
- Diffuse ONH edema that may be unilateral or bilateral
- Relatively mild vision loss
- No altitudinal defect on VF; various patterns of mild loss seen
Diabetic Papillitis

- Slow resolution of ONH edema but complete or nearly complete recovery of vision is the norm
- Like NAION, more prominent in nerves with small cups
- Is it real.............or just a variant of NAION?
Grave’s disease

- Remember No SPECS........
- Soft tissue edema
- Proptosis
- EOM involvement
- Corneal involvement from exposure
- Sight threatening complications
- Hyper (most common), hypo, or euthyroid
Grave’s disease

- The sight threatening complication is optic neuropathy from compression at the muscle cone
- Requires oral steroids and/or orbital decompression
- Type II Grave’s patients
- 75-80% of Grave’s patients are smokers!
The end!