Papilledema vs. Pseudopapilledema

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Financial disclosures

- No financial disclosures
Examination Techniques

- Stereoscopic viewing essential
- VA and VF
- Spontaneous / elicited venous pulsation
- Pupil testing and color vision
- Brightness comparison and red cap test
Papilledema

- Bilateral* optic nerve head swelling secondary to increased intracranial pressure (always, by definition)
- Swollen, blurred margins with splinter hemorrhages and exudates as well as nerve fiber layer edema. Patton’s folds may be seen: concentric chorioretinal folds extending from the disc: only seen in papilledema
Papilledema

- May be asymmetric or very rarely unilateral (sequential swelling)
- VA varies but typically mild reduction only or no loss at all
- May get diplopia secondary to abducens nerve compression causing partial lateral rectus paralysis
- With increased ICP, can get choroidal folds only (before papilledema) at lower pressure levels
Papilledema

- VF usually shows an enlarged blind spot
- No pupillary defect. Normal color vision
- SVP / EVP absent with obliterated cup
Papilledema (IIH)
Papilledema IIH age 15
OCT
Papilledema (HTN)
Papilledema (tumor)
Subtle papilledema (IIH)
Papilledema IIH
Papilledema IIH
Papilledema IIH
Terson’s syndrome and papilledema

Due to subarachnoid hemorrhage traveling down optic nerve sheath
Papilledema progression
Patton’s Folds
Patton’s folds
Patton’s folds
Patton’s folds: RNFL thickness
231 in OD, 295 in OS
Patton’s folds: now you see them…….
Back then in 2007 you did not...
Longstanding papilledema with optic atrophy (IIH)
Papilledema OCT NFL
NFL edema
Papilledema OCT
Papilledema OCT
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 treatment.
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.
Increased ICP

- Studies show that ONH swelling as measured by OCT can decrease (but not instantly resolve) immediately after lumbar puncture.
- Measured in lateral decubitus position with OCT sideways!
- Shows that reduction of ONH compression is very rapid.
- Shows that pressure in spinal column is associated with pressure at ONH.
Etiologies of Increased ICP

- Space occupying lesion; must always be ruled out!
- Infection or anatomical abnormality
- Malignant hypertension
- IIH
- Certain medications
- 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)
- 5/100,000 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
- Emerging evidence that elevated testosterone / androgen levels may be the cause
- Sleep apnea link
- Can affect children, often overlooked
IIH

- Symptoms of transient blur, diplopia, tinnitus (intracranial noises, not just ringing), headaches, etc.
- ICP usually severely elevated; normal is 50 – 200 mmH20. Over 25 cm (250 mm) considered definitively abnormal. Single measurement can be misleading: levels can vary over 24 hours.
- Very rare variant of normal pressure IIH. S/S, but repeatedly normal ICP.
IIH

- Diagnosis requires normal MRI / MRV and CSF studies with elevated ICP
- Watch for spinal chord tumors
- Differential: Cerebral Venous Sinus Thrombosis
- MRV
CVST

- Mostly young women
- Often not overweight
- Can be life threatening
- Treat with blood thinners, Diamox

- Can be seen with MRI, but potentially missed if MRV not performed (MRV by far the most sensitive)
IIH Management

- Refer to a neurologist
- Medical management includes Diamox, Lasix
- Weight loss
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.
Chronic IIH induced edema leading to atrophy: S/P decompression

22 year old AA F

Light perception 10/700
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
Sphenoid wing meningioma
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
ONH Drusen
ONH Drusen
ONH Drusen
ONH Drusen
ONH Drusen
ONH drusen
ONH DRUSEN SD-OCT
ONH DRUSEN SD OCT
Color SD-OCT
ONH drusen detection with OCT

- Optic Disc Drusen Consortium Consensus......
- Always use EDI
- Blood vessels are more solid, cast a shadow, and can show as figure 8
- Drusen always prelaminar
- Drusen always hyporeflective
- Drusen often have a hyperreflective border, especially superiorly
ONH drusen detection with OCT

- Drusen can conglomerate, and these areas can have some internal reflectivity from borders.

- The old concept of a hypoflective fluid wedge at the edge of the nerve in true papilledema DOES NOT APPLY with SD-OCT. Was a time domain OCT artifact.
FAF ONH Drusen
FAF ONH Drusen
NFL loss with ONH drusen
IIH with ONHD and papilledema
IIH with ONHD and papilledema
ONH drusen MRI
ONH drusen B-scan
The end!