Papilledema vs. pseudopapilledema: Are they swollen or are they not?

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Financial disclosures

- No financial disclosures
Examination Techniques

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

- Bilateral (but can be sequential with one nerve becoming swollen before the other, thus unilateral at presentation) 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
Papilledema

- May be asymmetric
- 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 (IIH)
Papilledema IIH age 15
OCT
Papilledema (HTN)
Papilledema (tumor)
Subtle papilledema (IIH)
Papilledema IIH
Papilledema IIH
Papilledema IIH
Papilledema with Patton’s folds
Terson’s and papilledema
Papilledema progression
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...
Patton’s folds
Longstanding papilledema with optic atrophy (IIH)
Papilledema OCT NFL

<table>
<thead>
<tr>
<th>Normal distribution</th>
<th>Percentiles</th>
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<tr>
<td>Avg Diam</td>
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Signature: NFL edna 04/20

Physician: [Signature]
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 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.
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)
- 1/100,000 in population as a whole; 20/100,000 in 20 to 44-year-old women 10% over ideal weight
- May be related to medications including TCN (minocycline especially), 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
- Doubles cardiovascular risk in females
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 more rare over age 50

- Less often female
- Fewer headache complaints
- More frequently discovered incidentally due to papilledema with no symptoms
- Lower opening CSF
- More likely to have concomitant medical conditions
- Less likely to use tetracycline family antibiotics
IIH

- Diagnosis involves normal MRI / MRV and CSF studies with elevated ICP
- Watch for spinal chord tumors
- Differential: Cerebral Venous Sinus Thrombosis
- MRV
CVST (cerebral venous sinus thrombosis)

- Young women and some men
- Often not overweight
- Can be life threatening
- Treat with blood thinners, Diamox

- Can be seen with MRI, but potentially missed if MRV not performed
- Stenosis may be secondary to IIH
Optic atrophy post CVST induced papilledema
IIH Management

- Refer to a neurologist
- Medical management includes Diamox, Lasix, Topamax
- 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
Papilledema IIH opening LP 550
After 3 weeks on Diamox
Side by side comparison
Minocycline induced elevated ICP papilledema
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 hyper-reflective border, especially superiorly
Drusen can conglomerate, and these areas can have some internal reflectivity from borders.

The old concept of a hypo-reflective fluid wedge at the edge of the nerve in true papilledema DOES NOT APPLY with SD-OCT. Was a time domain OCT artifact.
Peripapillary Hyper-reflective Ovoid Mass-like structures (PHOMS)

- “Fomms”
- Seen best with EDI
- Only seen with OCT, nothing else
- Circular innertube like structure around the disc above Bruch’s membrane
- Herniated optic nerve fibers
- Seen in any condition that leads to nerve swelling or congestion
- ION, papilledema, disc drusen
FAF ONH Drusen
FAF ONH Drusen
NFL loss with ONH drusen
Longstanding ONH drusen OU & new cat scratch disease OS
IIH with ONHD and papilledema
IIH with ONHD and papilledema
ONH drusen MRI
ONH drusen B-scan