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









## 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: RNFL thickness 231in 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



## NFL edema



## Papilledema OCT





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## Papilledema OCT





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- 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

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 values are one-way then the swelling will not improve rapidly with Tx

- 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

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.  $\diamond$  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



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#### 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



#### Light perception



#### 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 SD-OCT



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#### ONH DRUSEN SD OCT

## High Definition Images: HD 5 Line Raster OD 🔘 🔵 OS Scan Angle: 0° Spacing: 0.25 mm Length: 6 mm

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#### Color SD-OCT



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#### 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

#### ONH drusen detection with OCT

 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) Herniated optic "Fomms" nerve fibers Seen best with EDI Seen in any Only seen with condition that OCT, nothing else leads to nerve Circular innertube swelling or like structure congestion around the disc ♦ ION, papilledema, above Bruch's disc drusen membrane



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#### 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**

