Papilledema vs. Pseudopapilledema

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The Normal Optic Disc

Cup-to-Disc Ratio (C/D)

• On average with Gaussian distribution and less than 5% with C/D = 0.7 or greater
• Physiologic cups tend to be symmetric between both eyes with an interocular C/D > 0.2 about 1% of the time
• Blacks have larger C/D than whites
• Cup size may increase slightly with age

Papilledema*: Definition
Optic disc edema secondary to increased intracranial pressure

* In the old days the optic disc was called the "papilla"; edema is the abnormal accumulation of fluid in tissues or body cavities; in other words, it is reflective of a disease process

Papilledema and Pseudopapilledema

• In both cases the optic disc is elevated
• In papilledema the nerve is elevated because it is swollen, or edematous because of increased intracranial pressure
• However, not every swollen or edematous optic nerve is the result of increased intracranial pressure
• In pseudopapilledema the nerve is elevated for structural reasons that do not involve swelling or edema and therefore is not necessarily reflective of a disease process

Optic Disc Edema w/o Increased ICP

• Optic neuritis (papillitis)
• AION
• Partial CRVO
• Infiltration of nerve: eg, sarcoid, leukemia
• Compression of intraorbital optic nerve: eg, ONSM, ONG
• Leber’s hereditary optic neuropathy
• Diabetic papillopathy
• Hypertensive disc edema
• Amiodarone optic neuropathy

Back to Papilledema…

Histopathology of Papilledema

Optic Nerve Orbital Anatomy

Retinal Fold
Subarachnoid Space
Displacement of photoreceptors

Optic nerve
Cranial nerves
Orbital artery
Orbital veins
Boehm’s sphincter
Inferior oblique
Superior oblique
Extraocular muscles
Cilia
Cornea
Sclera
Anterior chamber
Iris
Lens
Vitreous
Ciliary body
Spongiosa
Optic canal
Optic chiasm
Optic tract
Mammillary body
Habenular area
Diaphragm of the sella turcica
Cavernous sinus
Mechanisms of Papilledema

Frisen Grades of “Swelling of the Optic Nerve Head”

- **Grade 1**: C-shaped blurring of nasal, superior and inferior margins of disc; temporal margin normal
- **Grade 2**: 360-degree elevation of the disc margin
- **Grade 3**: Elevation of entire disc with partial obstruction of one or more of the retinal vessels at the disc margin
- **Grade 4**: Complete obliteration of cup and complete obstruction of at least some of the vessels on the surface of the disc
- **Grade 5**: Dome-shaped appearance with all vessels being obscured (“champagne cork” papilledema)

Most Common Cause of Papilledema in Ambulatory Population is Idiopathic Intracranial Hypertension (aka Pseudotumor Cerebri)*

- **Modified Dandy Criteria**
  - Presence of signs and symptoms of increased intracranial pressure
  - Absence of localizing neurologic findings except as they relate to elevated ICP
  - Normal mental status
  - Normal neuro-imaging (eg, normal ventricles, no evidence of brain tumor)
  - No other cause of increased intracranial pressure present

* But this must be a diagnosis of exclusion

Other Signs Consistent With Papilledema

- Flame shaped hemorrhages
- Paton’s lines
- Intraretinal exudates
- Intraretinal hemorrhages

Unilateral or Asymmetric Papilledema

In IIH about 10% of cases have asymmetric swelling - a difference of at least two grades of papilledema

How else can we evaluate the Optic Disc?

- Fundus Photography
- IVFA
- OCT
- FAF
- B-scan Ultrasound

IVFA in Papilledema

OCT in Papilledema

Despite high-grade papilledema for over 4 months, GCL+IPL thickness and visual fields are are preserved OU!
Signs of Chronic Papilledema

- Milky-gray color to optic disc
- Drusen-like changes within disc substance
- Drop-out of NFL
- Choroidal neovascular membranes (rare)

Postpapilledema Optic Atrophy

- Takes weeks to develop
- Attenuation of retinal vessels
- Shunt vessel formation
- Peripheral nerve fibers drop out before papillomacular bundle fibers

Visual Field Defects in Papilledema from Idiopathic Intracranial Hypertension (Pseudotumor Cerebri)

- Enlarged Blind Spot
- Diffuse Constriction
- Inferonasal Step
- Arcuate Defects
- Central Scotomas

Pseudopapilledema vs. True Papilledema

<table>
<thead>
<tr>
<th>Finding</th>
<th>Pseudo</th>
<th>True</th>
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<tbody>
<tr>
<td>SVP</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Hemorrhages</td>
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<td>Yes</td>
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<tr>
<td>Retinal Striae</td>
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<td>Yes</td>
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<tr>
<td>Enlarged BS</td>
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<td>Yes</td>
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<td>TVO</td>
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<td>FluoroLeakage</td>
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<td>Other Neuro</td>
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<td>Yes</td>
</tr>
<tr>
<td>High Hypermet</td>
<td>Yes</td>
<td>No</td>
</tr>
</tbody>
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Ophthalmoscopic Features of Pseudopapilledema

- Elevated disc; margins obscured
- Absence of physiologic cup
- Vascular anomalies with increased branching
- Normal nerve fiber layer; disc transilluminates
- May have spontaneous venous pulse
- May see disc drusen (“hyaline bodies”)
- Hemorrhages rare
- No exudates
- No nerve fiber layer infarcts

Pseudopapilledema

- Structural congestion
- Drusen of optic disc
- Tilted optic discs
- Dragged disc (in ROP)
- Myelinated nerve fibers
- Glial veil
- Dysplastic optic discs, e.g.,
  - Megalopapilla
  - Morning Glory syndrome
Pseudopapilledema

- Structural congestion

Pseudopapilledema with Structural Congestion

“Little Red Discs”

Pseudopapilledema with Segmental Elevation

Pseudopapilledema with Tortuous Retinal Vessels

Midline Facial Defects are often Associated With Dysplastic Optic Nerves

Pseudopapilledema with Anomalous Retinal Vessels

Pseudopapilledema with Tilted Disc
Pseudopapilledema with “Dragged” Disc

Myelinated Nerve Fibers

Glial Veil

Drusen
- Druse/Drusen - German for crystals
- Two types: macular drusen and optic disc drusen
- Are not the same histologically
- Buried optic disc drusen
  - Look at parents’ optic discs
  - Use red-free light on ophthalmoscope

Visible Optic Disc Drusen

ON Drusen and Visual Field Defects
- ON Drusen can cause visual field defects that are indistinguishable from glaucoma

Pseudopapilledema with Buried Drusen

Abnormal RNFL thickness is probably an artifact

Optic Nerve Drusen Visible on CT scan

Fundus autofluorescence shows abnormalities consistent with optic disc drusen
Disc Drusen Associated CNVM - After Laser Treatment

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- In pseudopapilledema the nerve is elevated for structural reasons that do not involve swelling or edema and therefore is not necessarily reflective of a disease process.
- Current ophthalmic imaging techniques are often crucial for the differentiation of these two conditions.