An Approach to Papilledema

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Learning objectives

1. Distinguish true papilledema from pseudopapilledema using examination and ancillary testing methods
2. Differentiate optic disc swelling from elevated intracranial pressure from other disorders
3. Initiate a directed and timely workup for underlying causes of papilledema
4. Institute a management plan based on best available evidence
Ancillary testing

Visual fields
- Automated perimetry
- Goldmann (kinetic) perimetry

Echography
- B-scan
- A-scan + B-scan

Optical coherence tomography
- Peripapillary retinal nerve fibre layer
- Macular GCIPL complex
- OCT-A
Papilledema vs disc swelling from other causes

Severity of vision loss vs disc swelling
Accompanying symptoms
Timing/onset of findings

Unilateral vs bilateral?
Presence of hemorrhages?
Cotton wool spots?
Demographics?
Idiopathic Intracranial Hypertension (IIH)

AKA pseudotumor cerebri
Elevated intracranial pressure without identifiable anatomic cause
Diagnosis generally conforms to modified Dandy criteria
Elevated ICP
Normal CSF contents
No focal neurological deficits (except...)
Normal neuroimaging
All criteria must be satisfied!!
Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children

<table>
<thead>
<tr>
<th>1. Required for diagnosis of pseudotumor cerebri syndrome(^a)</th>
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<tr>
<td>A. Papilledema</td>
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<td>B. Normal neurologic examination except for cranial nerve abnormalities</td>
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<td>C. Neuroimaging: Normal brain parenchyma without evidence of hydrocephalus, mass, or structural lesion and no abnormal meningeal enhancement on MRI, with and without gadolinium, for typical patients (female and obese), and MRI, with and without gadolinium, and magnetic resonance venography for others; if MRI is unavailable or contraindicated, contrast-enhanced CT may be used</td>
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<td>D. Normal CSF composition</td>
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<td>E. Elevated lumbar puncture opening pressure (≥250 mm CSF in adults and ≥280 mm CSF in children [250 mm CSF if the child is not sedated and not obese]) in a properly performed lumbar puncture</td>
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Findings

Focal stenosis

Smooth tapering stenosis or hypoplasia

Focal stenosis
IIH Treatment Trial

Double-masked, randomized placebo controlled trial of IIH treatment

- Mild pseudotumor cerebri
  - $2 \text{ dB} < \text{MD} < 7 \text{ dB}$
- Study drug up to 4 g/day
- Outcomes compared at 1 yr, longterm followup to 5 years
Change in Papilledema Grade
Worse Eye

Grades of Papilledema

0 1 2 3 4 5

p < 0.001
Acetazolamide combined with weight loss is more effective than weight loss alone
Effect on papilledema and quality of life
More severe vision loss/papilledema may benefit even more
Does not address worse than -7 dB!!
Severe papilledema is an emergency.

Pattern Deviation not shown for severely depressed fields. Refer to Total Deviation.

Not seen
MD: -34.49 dB
FL: 0/11
PSD: 1.46 dB
FN: -100%
FP: 0%

Pattern Deviation not shown for severely depressed fields. Refer to Total Deviation.

Seen
MD: -20.43 dB
FL: 1/18
PSD: 10.52 dB
FN: 0%
FP: 0%
Conclusions

1. Use appropriate testing methods to diagnose papilledema
   • Misdiagnosis is frequent
   • Can lead to unnecessary workup and treatment

2. Obtain timely imaging and lumbar puncture

3. Exclude secondary cause of papilledema/high ICP

4. Achieve long-term remission of disease