PITFALLS IN PAPILLOEDEMA

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Why care about papilloedema?

“Dr. the good news is that it’s not my problem.”

Does this disc demonstrate...

a) A life-threatening illness?

b) A physiological variant of no significance?

What is “papilloedema”?

• Syndrome of optic disc oedema secondary to ↑ ICP
• Features:
  - Bilateral (can be asymmetric)
  - Vision relatively preserved
• Significance
  - Indicates elevated ICP - ? cause
  - Vision potentially threatened

Intracranial pressure (ICP)
LM findings in papilloedema

- Optic disc oedema
- Swollen/fragmented prelaminar axon
- Dilated prelaminar CRV
- Collapsed postlaminar CRV

“CRVO” theory of papilloedema

EM findings in papilloedema

- Oedematous axons
- Axoplasmic stasis

Mechanical vs Ischaemic Theory

- Normal
  - Ciliary circulation shared by choroid and laminar region
- Mechanical
  - Raised CSF pressure distorts axoplasmic flow at lamina
- Ischaemic
  - Raised CSF pressure causes “choroidal steal” of ciliary supply

What are symptoms of raised ICP?

- Raised ICP
  - Headaches
  - Worse recumbent, AM
  - Associated nausea and vomiting
  - Pulsatile tinnitus
  - CSF leak
  - Neurological deficit
    - Non-localising
    - CN VI palsy
    - (Localising)
- Papilloedema
  - Hypermetropic shift
  - Enlarged blind spot
  - Transient obscurations of vision (TOV)
  - Positive visual phenomena
  - Decreased vision

Papilloedema - signs

- VA
- Colour vision
- **VF
  - Enlarged blind spot
  - Generalised constriction
- Pupils
  - RAPD if asymmetric
Modified Frisén scale

<table>
<thead>
<tr>
<th>Grade 0 (Normal)</th>
<th>Grade 1 (Minimal)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Discernible vein*</td>
<td>Disk/halo visible.</td>
</tr>
<tr>
<td>Grade 2 (Low)</td>
<td>Grade 3 (Moderate)</td>
</tr>
<tr>
<td>Discernible vein</td>
<td>Discernible leak.</td>
</tr>
<tr>
<td>Grade 4 (Marked)</td>
<td>Grade 5 (Severe)</td>
</tr>
<tr>
<td>Total obscuration in the disc of a segment of a major blood vessel on the disc.</td>
<td></td>
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</tbody>
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Other fundus findings

- Peripapillary haemorrhages
- Terson’s syndrome
- Retinal / choroidal folds
- Macular star / hemi-macular star
- Absence of spontaneous venous pulsations
- SVP present 85% normal individuals
- Presence makes ICP unlikely
- Look carefully!
  - Red-free filter on direct ophthalmoscopy
  - Ensure patient not breath-holding
- Explanation of SVP's (in 85% of normals)
  - Blood flows high pressure -> low pressure
  - Intra-ocular CRV pressure > Retrolaminar CRV pressure
  - Intra-ocular CRV pressure subject to IOP
  - Retrolaminar CRV pressure subject to ICP
  - IOP pulse pressure is 3 mmHg
  - CSF pulse pressure is 1 mmHg
  - During systole, intra-ocular CRV pressure rises 1mmHg more than retrolaminar pressure, so vein empties and collapses
  - During diastole, intra-ocular CRV pressure lowers 1mmHg less than retrolaminar pressure, so vein distends and fills
  - Elevated ICP (>20 cmH2O) eliminates this disparity in pulse pressure
Management of papilloedema

- Investigations
  - MRI/MRV brain
  - LP (unless SOL)
- Mx of underlying cause
- MX of ICP
  - Temporary – LP, lumbar drain
  - Medical – acetazolamide, topiramate
  - Surgical – LPS/VPS, ONSFS

IIH

- Aka BIH, PTC
- Overweight females of childbearing years
- Papilloedema
- Raised ICP
- Normal imaging and CSF constituents
- Generally “benign”
- Can have fulminant and slowly progressive forms
- Rx – weight loss, medical, surgical
- ? CVS stenting

Pitfalls in Papilloedema

Case 1

- Pt in 40’s
- Migraines
  - Headache now side-locked
  - Increased frequency and severity
- Binocular diplopia
  - Vertical / torsional
  - Greatest in left gaze
  - Worse with right head tilt
- Reduced vision
Case 1 (cont)

- Vision problems both eyes
- Needs to wear SVN Rx to see at distance
- Misses temporal letters
- Enlarged blind spots on confrontation VF

Pitfall #1 – not even looking in the eyes!

THM

- Ophthalmoscopy not widely or confidently performed in general medical circles
- Despite the fact that it is often indicated – eg headache, vision loss
- Surrogates for Ophthalmoscopy – photography, OCT
- While an MRI or CT might show signs of raised ICP, papilloedema itself will never be detected without fundoscopy!
- Important for:
  - Preventing vision loss
  - Monitoring ICP

Case 2

- 38 yo man
- Previously fit and well
- Blurred vision, headache for 1.5 weeks
- VA R 6/7.5, L 6/12
- PERRL nil RAPD
- “Bilateral Neuroretinitis”

Diagnosis?

BP 210/145

Pitfall #2 – Malignant Hypertension

- Clues on examination
- Always check BP first
- Optometrists can do this!
Case 3
- 22 yo female
- Presents headache, blurred vision, papilloedema
- Opening pressure 40 cmH2O
- Rx – VP shunt
- Complicated infection – removed
- DVT – warfarin
- IIH managed acetazolamide and topiramate

Case 3
- Ongoing difficulties
- Headaches persistent
- Worsening vision - VA to CF 0.5 m OU
- Poor response to medication
- Investigations
  - Repeat LP – OP 21 cm H2O
  - MRI – NAD
- Is shunt re-insertion required?

Case 3 - VF

Case 3 – disc OD

Case 3 – disc OS

Case 3 - OCT
Goldmann VF

Pitfalls in chronic management

- In patients with chronic raised ICP:
  - Headache symptoms may become more "generic"
  - Other headache types may be present – eg migraine, medication overuse
  - Imaging may not demonstrate hydrocephalus
  - "stiff-walled ventricles"
  - Disc oedema may not be present in setting of optic atrophy or ONSEFS
  - "Dead discs don’t swell"
  - Shunts frequently fail over time
- An OP at lumbar puncture is a "snapshot" in time
  - ? Peaks/troughs
  - False high readings are common – anxiety/tension
- High prevalence of migraine and functional vision loss in IIH
  - Solution – comprehensive assessment, appreciate limitations

Case 4

- 58yo man NSCLC metastatic
  - Rx erlotinib (tyrosine kinase inhibitor)
  - Cx acne – Rx minocycline
  - 6 months later
    - Headache, dizziness, pulsatile tinnitus
    - MRI brain normal
    - MRI spine 2 small leptomeningeal mets
    - LP – OP 25 cmH20, +ve cytology NSCLC
  - Rx for CNS mets – high dose erlotinib
  - 1 month later worsened symptoms
  - S/B optometrist – concerned papilloedema

Case 4 (Jan)

- Neurological and ophthalmological opinions – NAD
  - Minocycline ceased
  - ? Partial seizures
  - ? Vestibular dysfunction
  - Rpt MRI brain normal
  - Rpt LP 24 cmH20, normal CSF
  - Insistence from optometrist – commenced acetazolamide
  - Initial improvement in symptoms
  - Ultimately intolerant
  - Trialled frusemide, topiramate with little success
  - Rpt MRI brain again normal

Case 4 (March)
Case 4

- Symptoms continued to progress
  - Despite "normal" investigations
    - Persistence of papilloedema
    - VA 6/12 OU
  - Admitted for continuous ICP monitoring
    - Initial pressure 70 cmH20
    - Further pressure spikes to 40 cmH20
    - Immediate relief of symptoms
    - Unfortunately permanent Vision loss

Pitfall #4

- Papilloedema (and raised ICP) may need treatment to prevent permanent vision loss
  - In this case, the key finding was persistent papilloedema and symptoms
    - Negative or borderline investigations were misleading
    - Opening Pressure on LP is only a "snapshot" of ICP
    - Difficult cases may require ICP monitoring or empiric treatment

Case 5

- 16 yo overweight girl
- Headaches
- Transient visual disturbances
- Optometrist noted papilloedema

Case 5

- MRI Brain + MRV
  - Normal
- LP
  - OP 12 cm H20
  - Normal CSF
- Post LP headache
  - Epidural blood patch

Case 5

- Features of headache
  - Unilateral (either side)
  - Throbbing
  - Severe, disabling
  - Photophobia, phonophobia
  - Nausea ++
  - No postural relationship
Case 5 (cont)

Pitfall # 5 - Pseudopapilloedema

- Asymptomatic
- Cup absent
- Anomalous vascular branching
- Normal colour
- Irregular elevation
- Retinal veins not dilated
- No exudates
- SVP present

- Symptomatic
- Cup present
- Vessels normal
- Hyperaemia
- Diffuse elevation
- Retinal veins dilated
- Exudates
- SVP absent

SUSPECTED PAPILLOEDEMA:
A suggested approach for optometrists

- History and examination
  - Visual Fields (at least confrontation)
  - Colour vision (Ishihara plates each eye)
  - Pupils
  - Motility assessment
  - DFE and fundus lens assessment
  - Blood pressure

Category 1: “Urgent” papilloedema

- “The tempo of the presentation dictates the tempo of management”
- Concerns because:
  - Rapid onset of symptoms (esp. vision loss)
  - Severe vision loss
  - Focal neurological signs (eg. hemiparesis, dysphasia)
  - Severe disc oedema (florid papilloedema and haemorrhages, ischaemic changes, macular star)
  - Malignant hypertension
- Management
  - Risk of life-threatening disease (SOL, malignant HT)
  - Risk of irreversible blindness
  - Need urgent (same day) investigations and management
  - Hospital (TED – Neurosurgery/Neurology/Ophthalmology)

Category 2: “Genuine” papilloedema

- Definite symptoms and disc oedema
- Vision stable (enlarged BS OK)
- Chronic symptoms (months)
- Moderate papilloedema, no critical features
- Clinical suspicion of IIH

- Management:
  - “Prompt” outpatient referral to an appropriate specialist
  - Requires imaging, LP, etc. – as an outpatient

Category 3: It might be papilloedema…

- Asymptomatic
- Or symptoms otherwise explainable
- Asthenopia, migraine etc
- Normal vision
- Suspected pseudopapilloedema
- But you don’t want to miss something…

- Management:
  - Routine referral to appropriate specialist
  - Honestly explain to patient
  - Avoid catastrophising!
Thank You!

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