Papilledema & Idiopathic Intracranial Hypertension

Monterey Symposium
California Optometric Association 2019

Leonard V. Messner, OD, FAAO
Professor of Optometry
Vice President for Strategy & Institutional Advancement
Illinois College of Optometry

Financial Disclosure

- King Devick Technologies (scientific advisory board)
- Heidelberg Engineering (scientific advisory board)

34 y/o Woman

- VA = 20/20, OU
- + HA x 2 yrs.
- Normal neuro exam
- BMI: 39

IIH - Key Points

- Defining pseudotumor cerebri & IIH
- Pathogenesis of IIH
- Clinical features and neuroradiology
- Prognostic factors and management
- OCT analysis of papilledema
- Visual fields!

Dandy WE Ann Surg 1937
Revised Diagnostic Criteria for the Pseudotumor Cerebri Syndrome in Adults and Children

1. Signs and symptoms of increased intracranial pressure
2. Absence of localizing findings on neurologic examination
3. Absence of deformity, displacement, or obstruction of the ventricular system and otherwise normal neuroimaging studies, verified by evidence of increased CSF pressure ≥200 mm water determined neuroimaging studies for empty sella, or opacification of the sphenoid sinus and ethmoid recess on contrast-enhanced CT or MRI. Headaches and smooth swelling of the retouched visual fields or oculomotor should last for another diagnosis.
4. Positive and acute
5. No other cause of increased intracranial pressure present

For CSF opening pressure of ≥200 mm water: at least one of the following

- Pulsed synchronous limbic
- Test [date]
- Test [date] and [date]

Neuroradiology Findings with IIH

- Empty sella
- Posterior scleral flattening
- Optic nerve distension
- Ectopic displacement of cerebellar tonsils
- Dural venous sinus stenosis

Neuroradiology Findings with IIH

- Empty sella
- Posterior scleral flattening
- Optic nerve distension
- Ectopic displacement of cerebellar tonsils
- Dural venous sinus stenosis

Neuroradiology Findings with IIH

- Empty sella
- Posterior scleral flattening
- Optic nerve distension
- Ectopic displacement of cerebellar tonsils
- Dural venous sinus stenosis
Neuroradiology Findings with IIH

- Empty sella
- Posterior scleral flattening
- Optic nerve distension
- Ectopic displacement of cerebellar tonsils (21%)
- Dural venous sinus stenosis


Neuroradiology Findings with IIH

- Empty sella
- Posterior scleral flattening
- Optic nerve distension
- Ectopic displacement of cerebellar tonsils
- Dural venous sinus stenosis (90%)


27 y/o AA Woman

- c/o moderate, progressive headaches x 1-2 month
- Normal neurologic exam
- BVA:
  - 20/20 OD
  - 20/20 OS
- BMI: 58
Epidemiology of IIH

- 85 - 90% female
- 15 to 44 years
- Obesity
- MAI = 0.9-1.7/100,000
  - 7.9-20.2/100,000 if obese female 15-44 yrs.


Pathogenesis of IIH (???)

- Increased cerebral blood volume - Raichle’ 1978
- Increased arachnoid resistance to CSF drainage - Aisenberg’ 1980
- Increased intra-abdominal pressure with cerebral venous hypertension - Sugerman’ 2001
- Reduced CSF absorption by extracranial lymphatics - Johnston’ 2006

Baseline Clinical Profile (IIHTT)

- Headache (84%)
- Back pain (53%)
- Tinnitus (52%)
- Vision loss
  - Transient vision loss (68%)
  - Symptomatic vision loss (32%)
  - Visual fields (86% {5% > 15db})
    - Enlarged blind spot
    - Inferior arcuate defects
The rest of the story...

20/20

20/20

12 mos later...

-11.13 db

-3.22 db

19 mos later...

-23.54 db

-29.62 db

Risk Factors for Poor Visual Prognosis

• Male gender
• African American
• Morbid obesity
• Marked weight
• Obstructive sleep apnea
• Concomitant medications
• Acute onset of signs and symptoms of elevated ICP (fulminant IIH)
• High-grade papilledema
• Lack of headaches
• Lack of ophthalmic oversight

Biousse V. NANOS 2013
Gender

- Men represent approx 10% of all IIH
- Less likely to have headache ("estrogen factor")
- Less likely to seek care

Digre & Corbett Arch Neurol 1989
Bruce BB, et al. Neurology 2009

Race

- No strong influence on development of IIH
- African Americans = more aggressive disease:
  - 3X more likely to have severe vision loss
  - 5X more likely to develop blindness


Obesity

- NORDIC ave. BMI = 39
- Increased severity of papilledema and visual loss if BMI ≥ 40
- Recent weight gain (5-15%) even in non-obese patients

Wall & George. Brain 1991

Medications

- Many meds proposed to induce IIH, few test-retest data
- Tetracycline derivatives (minocycline)
- cyclosporine, lithium, nalidixic acid, nitrofurantoin, oral contraceptives, levonorgestrel, danaxol, and tamoxifen

Friedman DI. Am J Clin Dermatol 2005

14 y/o Girl

- HAs x 2-3 weeks
- Rx minocycline x 1 month for acne
- VA: 20/20 OU
- BMI: 23

31 32 33 34 35 36
• MRI / MRV normal
• D/C minocycline
• Follow-up x 3 weeks...

By 4 mos:
Reduction in RNFL:
109 microns OD
196 microns OS

Reduction in RNFL:
32 microns OD
142 microns OS
Co-Morbidities

- Anemia
- Hypertension
- Obstructive sleep apnea

Other Clinical Findings Predictive of Poor Outcomes

- Absence of headaches
- Severity of papilledema (+/-)
- Marked visual field defects at presentation
- “Fulminant” course (malignant IIH)

OCT Analysis of Papilledema

- Increased NFL thickness:
  - Nasal > temporal (nasal thickness > 78.0 microns)
- Elevation of nerve head (>0.8 mm from RPE to apex)
- Maintenance of central cup (until late disease)
- Subretinal hyporeflective space between photoreceptor layer and RPE (recumbent “lazy V”)  
  - Inward deflection of RPE/BM (N>T)
    - 67% with papilledema

32 y/o AA Woman

- C/o progressive headaches am > pm
- BMI: 41
- BVA:
  - 20/20 OD
  - 20/20 OS
37 y/o AA Woman

- C/o chronic daily headaches
- + synchronous pulsatile tinnitus
- BMI: 38
- BVA:
  - 20/20 OD
  - 20/20 OS
40 y/o AA Woman

- C/o chronic daily Has
- + SPT
- BMI = 44
- BVA:
  - 20/40 OD
  - 20/40 OS
38 y/o AA Woman

- Medical history = SLE
- Tx with po prednisone
- Approx. 200 lb weight gain (>400 lbs.)
- BVA:
  - 20/20 OD
  - 20/20 OS
Cont.

• MRI / MRV consistent with IIH
• Coordination of care through HMO PCP
  – Nothing done
• F/U @ IEI 5 mos. later complaining of progressive severe HA (10/10)
• BVA:
  – 20/20 OD
  – 20/20 OS

Management of IIH

• Observation
• CAI’s / diuretics
  – ? value
• Surgery
  – VP shunt
  – ONSF
• Weight loss
  – 5-10% BMI

“Visual Fields”
NORDIC IIH Treatment Trial

• Specific Aim 1: The trial is focused on determining the efficacy of low sodium diet with or without acetazolamide to reduce or reverse visual loss.

• Specific Aim 2: (a) To identify proteomic and genetic risk factors for IIH by screening a large cohort of IIH patients and controls.

IIHTT 6-Month Results

• Acetazolamide better than diet alone for visual field improvement (PMD reduction of 1.43 vs. 0.71 db)

• Acetazolamide arm showed better weight reduction than diet alone

• Acetazolamide arm showed better resolution of papilledema & improvement of QOL scores vs. diet alone


32 y/o Hispanic Woman

• c/o progressive, debilitating headaches x 2 mos.

• Normal neurologic exam

• BVA:
  – 20/20 OD
  – 20/20 OS

• BMI: 38
F/U x 6 mos

- Rx Diamox (500 mg BID)
- Weight loss (approx. 25 lbs.)
- Improvement in headaches
F/U x 14 mos

- D/C Diamox x 3 months
- Weight loss (BMI reduction from 38 to 30)
- Headache free
IIHTT Visual Field Performance Failures

- PF was identified in 21% of participants and in 2.7% of the total number of VF examinations and was reversible on repeat testing
- When perimetric worsening appears to have occurred in someone with papilledema who otherwise is clinically stable or improving, retesting is likely to reveal that the apparent worsening is due to poor performance rather than true worsening of the condition


But what if I still have a headache?

Headache with IIH

- Most common symptom with IIH (84% in IIHTT)
- 41% with prior history of migraine
- 2/3 persistent HA after remission of IIH (improvement of papilledema)
  - Sensitization of peripheral & central neurons over time

Friedman DI. NANOS 2019

IIH - Key Points

- Defining pseudotumor cerebri & IIH
- Pathogenesis of IIH
- Clinical features and neuroradiology
- Prognostic factors and management
- OCT analysis of papilledema
- Visual fields!

Thank you!