Intracranial Hypertension (Pseudotumor cerebri)

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Introduction

IIH is a condition marked by:
- Unknown cause
- Elevated cerebrovascular fluid (CSF) pressure
- Papilledema

Also referred to as
- Pseudotumor cerebri syndrome
- Benign intracranial hypertension

Incidence

- General population: 1:100,000
- Obese women of childbearing age: 19:100,000
- Can develop in children
  - Male = female
  - Typically precipitated by antibiotic or steroid withdrawal
Age at Diagnosis

Major Symptoms

- Headache
  - Worse in AM
  - Exacerbated by Valsalva maneuver
- Transient visual obscurations
  - 1-5 sec. "graying-out"
  - Induced by orthostatic changes
- Horizontal diplopia
- Sixth nerve Palsy

Less common symptoms

- Pulsatile tinnitus
- Pain associated with
  - Neck
  - Shoulders
  - Back
Symptoms

Consciousness is Never Altered

Physical signs

- Cardinal sign is Papilledema
  - Usually bilateral
  - May be asymmetrical
  - Occasionally unilateral

ONH
Permanent Sequelae

- Visual loss due to papilledema
- Occurs in 10-25% of patients
- Including children!

Diagnostic methods

1st step is to Rule Out:
- Brain tumor
- Hydrocephalus
- Infections
- Other treatable diagnostic possibilities
- Exogenous substances
- Systolic Disease
- Disorders of cerebral venous drainage
Diagnostic methods

• 1st step is to Rule Out:
  • Brain tumor
  • Hydrocephalus
  • Infections
  • Other treatable diagnostic possibilities

USE CAUTION

• When diagnosing IIH in
  • Men
  • Non-obese females
  • Children
  • Pts. W/ HA & Papilledema but normal CSF Pressure

Vision Assessment

• Visual Acuity (VA)
• √ RAPD
• EOMs
• Visual Field Testing
• Fundus Photography
• ?Visual Evoked Potential (VEP)
Lab Studies

- Lumbar puncture (LP) is **MANDATORY**
- CSF ≥ 250 mm H₂O
- CSF cytology to exclude:
  - Inflammation
  - Tumor cells
  - Infection

Typical CSF in IIH

- Normal or low protein (< 20 mg/%) 
- Normal glucose 
- Normal cell count

Additional lab

- ANA
- RPR, VDRL
- FTA-ABS, MHA-TP
- Serum Calcium
- (R/O hypoparathyroidism)
Endocrine studies

• Only if suspicion of
  • Hyperadrenalism (Cushing's)
  • Hypoadrenalism (Addison's)
  • Hypoparathyroidism

Imaging Techniques

• CT
• MRI
• DISA
• Arteriography

CT Findings

• Normal or small ventricles
• 25% - 50% of patients exhibit an empty sella
CT vs MRI

- Both effective to screen for other causes of intracranial hyper-tension such as:
  - Tumor
  - Hydrocephalus
  - Both can usually detect a venous sinus thrombosis

MRI

- More expensive
- Least invasive
  - for detecting venous sinus occlusions and tumors without IV contrast.
  - MRI may be impractical in
    - Obesity
    - Claustrophobia
  - DISA is occasionally more sensitive than MRI in detecting venous sinus obstruction.

Treatment

- Varies depending on:
  - Symptoms
  - Severity of vision loss
  - Presence of other conditions:
    - Glaucoma
    - Systemic hypertension
    - Renal disease
    - Systemic lupus-erythematosus
    - Pregnancy
Specific Treatment Modalities

- Diet
- Medication
  - Acetazolimide
  - Furosemide
- Repeated lumbar puncture
- Surgery

Diet

- Weight-reduction diet should be offered to all IIH pts.
- Pts. should be sent for professional dietary instruction.

Medication

- Headache
  - Beta-adrenergic blockers
  - Calcium channel blockers
  - Tricyclic antidepressants
- Visual Symptoms
  - Carbonic anhydrase inhibitors
  - Furosemide
  - Steroids
Repeated Lumbar Puncture

- Advocated by some
- No clear correlation between severity of HA and height of CSF elevation.
- Drawbacks:
  - Painful
  - Difficult to perform in obese pts.
  - Post-LP HA

Surgery

- Treatment of choice for pts. who:
  - Are losing vision
  - Have unremitting HA unresponsive to medications
- Surgical options:
  - Optic Nerve Sheath Fenestration
  - Lumboperitoneal Shunt
  - Ventriculoperitoneal Shunt

Optic Nerve Sheath Fenestration (ONSF)

- Preferred operation
- Effectively preserves or restores vision in 80% - 90% of cases.
- Less reliable for relief of headache (60% - 65%).
Lumboperitoneal Shunts (LPS)
- Effective at relieving headaches
- Many complications
  - Can not be recommended for any patient who may require abdominal surgery
  - Propensity for delayed failure
    - Reoperation is the rule
    - Requires careful, longterm neurosurgical follow-up
  - Failure can lead to abrupt return of papilledema and vision failure.

Ventriculoperitoneal Shunt (VPS)
- Newer technique using stereotactic placement
- As effective as functional LP shunt
- Less complications?

Indications for Surgery
- Development of a new or enlargement of a pre-existing visual field defect.
- Presence of severe visual loss in one or both eyes at the time of 1st exam.
- Anticipated hypotension induced by treatment of high blood pressure or renal dialysis.
Indications for Surgery

- Psychosocial reasons such as pts. inability to perform VF testing, noncompliance with medical treatment, or an itinerant life-style.
- Headache unresponsive to standard headache regimens.

Specific Treatment Problems

- The Asymptomatic Patient
  - May not need Tx
  - Follow up q 1-3 months
- Headache
  - 
  - 
- Serious Visual Loss
  - ONSF
- Rapidly Progressive Visual Loss
  - ONSF + LPS/VPS + Meds + Beta-Blocker
- Pregnancy with Headaches
  - Beta-Blockers, LPS/VPS, bedrest, ONSF
- Renal Disease
  - ONSF
  - Beware hypotensive episodes

Prognosis

- Variable course
  - Most cases last 1 year or longer
  - Some may resolve in a few months
  - Some may linger for years
Management

Treatment of Idiopathic Intracranial Hypertension

- No visual loss
  1. weight loss
  2. symptomatic headache
  3. treatment
- Visual loss
  1. weight loss
  2. acetazolamide or furosemide
  3. progression

- Resolution
  - Steroids (2 mg x 2 mg, taper)
  - Half-closed tenectomy
  - Peritoneal or
  - Subcutaneous headache
- Lumbar punction