Spontaneous Intracranial Hypotension

Diagnosis and Treatment

John W. Engstrom MD, Philip R. Weinstein MD, and William P. Dillon M.D.
University of California, San Francisco

Spontaneous Intracranial Hypotension

• A spontaneous loss of CSF volume
  — resulting in orthostatic headache syndrome
  — usually postural in nature, occurring in all age groups, but often in young to middle-aged adults; females more common
• Usually due to a spinal CSF fistula at thoracic level or cervicothoracic junction; can occur at lumbosacral level
  — perineural (Tarlov’s) cyst or HNP/osteophyte that erodes dura
• CSF opening pressure is low, often below 6cm H₂O
  — But— pressures may fall within normal limits (6-20cm)
• Epidural blood patch is curative ~ 75% of cases
• Imaging is usually diagnostic, but can be subtle (MR>CT)
  — Slow CSF leak may result in “normal” CT myelogram
  — Fast leak may require C1-2 puncture in CT to localize site of leak

What’s inside the skull ...

• Brain ~ 1400 grams / 50 in water
• Vascular structures
• Cerebrospinal fluid
  — Intracranial volume ~150-200cc
  — Spinal volume 50cc
  — 0.3 ml/min in adults (500 ml/24h)
  — Opening pressure: 6-20 cm water (in lateral decubitus position)
  — Pressure at vertex when erect < 0cm

10% reduction volume = headache


Monro-Kellie Doctrine

The total volume within an intact skull is constant

The volume of brain, blood, and CSF are in reciprocal relationship

If one of these three elements increases (or decreases) in volume, this must occur at the expense of volume of the other two elements.
SIH: CLINICAL FEATURES

- **Headache**
  - Usually Postural, often abrupt in onset, variable
  - Headache becomes constant if chronic or if SDH occur
- **Neck Pain**: suboccipital, upper neck, upper extremities, interscapular
- **Visual**: Photophobia; Nystagmus
  - Horizontal diplopia (unilateral or bilateral 6th, 3rd nerve palsy)
  - Inferior Quadrantanopsia field defects (superior binasal field defects)
- **Auditory**:
  - Tinnitus (postural), vertigo (nausea), hyperacusis, hearing loss
- **Mentation**: memory loss, confusion, stupor, coma
- **Associated Syndromes**: Marfan’s, Ehlers-Danlos, NF1

Headache

- 15-30 min. after erect
- Exertional; worse at end of day
- Aggravated by cough, strain
- Paradoxical
- Mechanism
  - Traction on dura, CN’s (V)
  - Venous or sinus dilation

Complications of SIH

- Cerebral sinus thrombosis
- Pseudo-hypoxic cerebral edema
  - Thalamus, basal ganglia
- SDH/hygroma
- ICH
- Pneumocephalus (post-op)
- Dural-pleural fistula with pleural effusion
- Recurrent SIH new leak location: 5/13 = 38% within 3mos. after surg. repair T10-C7 (multiple cysts-3) (ct disorder-2)

SPONTANEOUS INTRACRANIAL HYPOTENSION: MR Findings

- Diffuse Dural Enhancement (95%)
- Brain Sag (80% severity)
  - Downward Displacement Of Pons, Cerebellum, Chiasm, Cisterns
- Venous Sinuses distention (80%)
  - Cranial and Spinal veins
- Pituitary Gland Prominence
- Subdural Fluid / Hematomas
- Midbrain compression from descending transtentorial herniation of temporal lobes

Schievink WI, JNS 2003 (99) 5
Mnemonic: SEEPS
- SDH
- Enhanced meninges
- Engorged veins
- Pituitary edema
- Sagging brain

SIH: VENOUS PROMINENCE

MR findings rapidly correct after treatment of SIH

Pre-therapy

Post-therapy

SIH: Dural enhancement
Reverts to normal following treatment

Venous Distension Sign
Farb et al
AJNR 28:1489, 2007
Spinal Manifestations of SIH

- Extradural collections of CSF
- Enlarged epidural veins, dural enhancement
- HNP/dural ectasia/root cysts
- Contrast at C1-2 (often false localizing)
- Siderosis (rare)
- Syrinx or Pre-syrinx secondary to tonsillar impaction

Burtis et al: Spinal manifestations of SIH AJNR 26:34-38, 2005

SIH Is Often Misdiagnosed

Wouter I. Schievink, MD Arch Neurol 2003;60:1713-1718

- 17 of 18 patients (94%) initially received an incorrect diagnosis: delay of diagnosis ranged 4 days to 13 years
  - Migraine, "meningitis," and psychogenic disorder
  - Subarachnoid Hemorrhage
- 30% underwent unnecessary procedures
  - Cerebral arteriography
  - Craniectomies for Chiari malformation (2), SDH (1), and dural and brain biopsy (1)

37-year-old previously healthy man with abrupt onset of headache not postural in nature

Initial CT at time of headache onset: Normal
Diagnosis in ED = "migraine"

Continues with headache over next 9 weeks, and re-presents to ER with postural headache, normal mentation
ER ordered CTA to rule out SAH

Tonsils low, "tight brain," bilateral subdural hematomas, but... prominent venous structures
Diagnosis suggested by radiologist, blood patch performed--more to tell...
Diagnosis: SIH  
Treatment: 20cc Epidural Patch  
Outcome: Resolution of HA 10 hours

9cc blood: 1 cc 240% omnipaque  
Slow injection, reverse trendelenberg  
Rotation on sides and back

Headache returns after three days

Thoracic blood patch, 20cc  
(Note the large transdural thoracic disc herniation  
Postural headaches resolve, but persistent headache requires subdural evacuation; resolution of symptoms

UCSF 2010 Review SIH  
Purpose and Methods

• Retrospective review of records of patients with the term “SIH” from 1995-2009  
• 73 patients identified  
• 37 patients had only MRI of the brain  
• 36 patients investigated with spine imaging  
• Noncontrast CT or MRI of the spine  
CT or MR Myelography

Results of those with Spine Imaging
Blood Patch Results

- 30 patients with SIH syndrome treated by early lumbar epidural blood patch (EBP) without previously performing lumbar puncture or identifying a CSF leak
- Complete cure was obtained in 77% of patients after one (57%) or two (20%) EBPs
  

Post-treatment Intracranial Hypertension

Intracranial hypertension after treatment of spontaneous cerebrospinal fluid leaks. Mokri B 2004

- 4 patients developed intracranial hypertension after treatment of their CSF leaks
- Headaches different from their previous headaches
- All had increased CSF opening pressure, bilateral papilledema (1), no venous pulsations (1)
- One responded to acetazolamide
- HA Gradually resolved in all cases.. ??? Increased CSF production

SUMMARY
SPONTANEOUS “CSF HYPOVOLEMIA”

- Often unrecognized cause of headache syndrome
  
  -- MR IS OFTEN DIAGNOSTIC, may localize; AND CAN LEAD TO THE CORRECT DIAGNOSIS AND THERAPY
- Epidural Blood patch is effective in over 75% of patients
  
  -- first line of therapy BEFORE myelogram evaluations or surgery
- Perineural cysts and transdural herniation of discs are frequent causes of leaks. Cord herniation may also occur
  
  -- Slow CSF leak may result in negative imaging studies
    • Delayed scans may help (1-2 hours)
    • Dynamic CT or Gad MR myelography may also show slow leaks
    • Isotope study rarely useful

All night: his finger in the dike
THANK YOU