First Annual Cedars-Sinai Intracranial Hypotension Symposium - October 14, 2017: Rebound Intracranial Hypertension - with Dr. Peter Kranz, Duke University Medical Center

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Rebound intracranial hypertension
March 1994
My first patient

John F. DOS: 3-12-1994
41 year-old man
8 year history of orthostatic headaches
MRI brain: brain sagging
CT-myelogram: single lumbar nerve root cyst
Treatment: surgery
Outcome: reverse orthostatic headaches 2nd postop day and visual loss after 6 weeks with papilledema and retinal hemorrhage
Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension

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W. L. Scheinok, et al.

Case 1. This man, initially presented at age 33 years with a 1-year history of progressive, daily occipital headaches. These headaches would invariably go away if he would lie down. A gastroenterological evaluation for associated nausea and vomiting was normal.

Examinations: General: Medical and neurological examinations were normal. Magnetic resonance imaging of the lumbar spine revealed a CSF leak at the L-2 level. CSF samples were obtained from the lumbar, thoracic, and cisternal spaces.

Laboratory: Protein in the cisternal fluid was elevated to 30 mg/dl with no evidence of traumatic xanthochromia. The protein in the lumbar and thoracic spaces was normal. The level of CSF in the lumbar space was normal, but the protein level was elevated to 22 mg/dl. The level of CSF in the thoracic space was normal, but the protein level was elevated to 30 mg/dl. The level of CSF in the cisternal space was normal, but the protein level was elevated to 30 mg/dl.

The patient was treated with corticosteroids but showed no improvement. Because of the patient’s persistent headaches, a decompression of the intracranial compartment was performed (Fig. 1).

The patient recovered well from the surgery and experienced a gradual improvement of the headaches for approximately 4 years, although they never resolved. He was able to lead a relatively normal lifestyle. However, the positional headaches worsened again over the ensuing 3 years. The headaches had been associated with nausea and vomiting, but now the patient also developed dizziness, dizziness, and bowel and bladder incontinence. Neurological examination showed generalized hyporeflexia. Magnetic resonance imaging revealed diffuse dural enhancement, displacement of the cerebellar tonsils to the level of C-3, elevation of the transverse sinus, and a small subdural hematoma at the level of the L-2 pedicle. A lumbar puncture made with the patient in the seated position showed an opening pressure of 6 cm H2O, total protein of 60 mg/dl, and low nucleated cell count. Bilateral injection of isotope-labeled human albumin (Fig. 2) with a radionuclide cisternography showed minimal migration of tracer over the cerebellum and a normal uptake of tracer in the upper lumbar spine region on the left (Fig. 3). Computed tomographic myelography was suggestive of a meningeal detachment at the level of the L-2 pedicle, indicating a long-standing process (Fig. 5). In retrospect, an abnormal CT scan performed 8 years previously for the evaluation of associated nausea and vomiting showed the presence of this detachment (Fig. 6).

Operation: A left L-2 hemilaminectomy and total facetectomy were performed. Extruded CSF was encountered immediately after removal of the ligamentum flavum. A meningeal dissection with multiple anastomoses was found at the L-2 nerve root. There was significant arachnoiditis and CSF leakage from the dura mater, which had entered the L-2 pedicle. The detachment was ligated circumferentially with several sutures without compromising the nerve root. There was no evidence of ongoing CSF leak.

Postoperative Course: The patient recovered well from surgery with complete resolution of all the complaints. Leg strength and sensation remained normal. One week after surgery, he noted occasional mild visual blurring. Six weeks postoperatively, examination showed bilateral papilledema with a homonymous inferior to the optic disc. The CSF protein was 40 mg/dl. Three months after surgery, he noted photophobia, and the papilledema resolved. Four months later the papilledema improved considerably and the homonymous field defect was resolved. Magnetic resonance imaging of the brain revealed resolution of the meningeal detachment at the level of the cerebellar tonsils, and normal improvement of the cerebellar displacement (Fig. 4).

We postulate that the development of papilledema is this patient may have been due to the sudden interruption of the abnormal pathway of CSF absorption, which had been present for many years.

The syndrome of spontaneous intracranial hypotension is characterized by a postural headache that may be associated with a variety of symptoms including proctor rhea, dizziness, nausea, vomiting, diplopia, visual blurring, tinnitus, vertigo, and a low back pain (1,2,3).

The diagnosis is confirmed by lumbar puncture, which reveals a low CSF pressure. However, variable modifications in CSF pressure occur at different positions, indicating that the CSF leak is intermittent. Examination of the CSF itself often shows reduced elevations of total protein and increased cell count. Characteristically, cranial MR imaging studies in pa...
Rebound intracranial hypertension
October 2017

• Routinely diagnosed?
Lack of knowledge of rebound high-pressure headaches

• Frequency: 0-20%
• Treatment: Acetazolamide and ....
• No diagnostic criteria
Rebound high-pressure headache after treatment of spontaneous intracranial hypotension. An MRV study

• 113 consecutive patients with SIH
• Diagnostic criteria for rebound headache
  a: reverse orthostatic headache
  b: resolution of headache after Diamox
  c: not better accounted for by another cause of headache
• MRV scores according to Higgins et al (JNNP 2004)
  0: normal
  1: one or more areas of focal narrowing
  2: one or more signal gaps
Rebound high-pressure headache after treatment of spontaneous intracranial hypotension. An MRV study

67 women / 46 men
Age: 45.9 (range, 13 – 71 years)

Rebound high-pressure headache: 31 (27.4%)

More common in
a) women (p=0.0474)
b) younger age (p=0.0135)
c) presence of extradural CSF (p=0.0286)
Rebound high-pressure headache after treatment of spontaneous intracranial hypotension. An MRV study

Frequency of rebound high-pressure headache

<table>
<thead>
<tr>
<th>MRV score</th>
<th>Count (n)</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>42</td>
<td>14%</td>
</tr>
<tr>
<td>1</td>
<td>34</td>
<td>24%</td>
</tr>
<tr>
<td>2</td>
<td>34</td>
<td>44%</td>
</tr>
<tr>
<td>3</td>
<td>3</td>
<td>67%</td>
</tr>
<tr>
<td>4</td>
<td>0</td>
<td></td>
</tr>
</tbody>
</table>

P=0.0092
Conclusions

• Rebound high-pressure headache is common after treatment for SIH (about one-fourth)

• Related to: Age, sex, presence of extra-dural CSF on spinal imaging, and venous anatomy on MRV
Rebound Intracranial Hypertension

Peter G. Kranz, MD
Duke University Medical Center

Disclosures

1. No conflict of interest
2. Use of fibrin glue for epidural injection is off label
RIH: What is it?

- New headache type after blood patching
- Characterized by:
  - HA worse when lying down
  - Often change in location – frontal
  - Nausea, blurry vision common
- Increased CSF pressure compared with baseline
  - May or may not be > 20 cm H₂O
- Onset immediately after up to days after
  - Usually worst 24-36 hrs post-patch

RIH: Examples

Initial pressure: **7 cm H₂O**

Overnight developed new sx
- non-positional HA
- Worse with lying down
- N/V, blurred vision

Lumbar puncture (+24 hr)
- Pressure: **28 cm H₂O**
- Drained to **9 cm H₂O**

Lumbar puncture (+48 hrs)
- Pressure: **29 cm H₂O**
- Drained to **7 cm H₂O**

**Brain MRI = SIH**

**Myelo = leak**

**Patch**

3 years prior

RIH: Examples

<table>
<thead>
<tr>
<th>Patient</th>
<th>Opening Pressure</th>
<th>Time from EBP</th>
<th>Headache Location</th>
<th>Post-EBP N/V</th>
<th>N/V improved</th>
<th>Post-EBP Blurred Vision</th>
<th>Post-EBP Blood Pressure</th>
<th>Patching Agent</th>
<th>Patch Volume</th>
<th>Duration of Acetazolamide Treatment</th>
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<tbody>
<tr>
<td>1</td>
<td>6.6</td>
<td>25.5</td>
<td>6 hours</td>
<td>Occipital</td>
<td>Yes</td>
<td>Frontal</td>
<td>Yes</td>
<td>Form glue</td>
<td>6 mL</td>
<td>5 days</td>
</tr>
<tr>
<td>2</td>
<td>N/A</td>
<td>28.0</td>
<td>3 weeks</td>
<td>Retro-orbital</td>
<td>No</td>
<td>No</td>
<td>Blood</td>
<td>Blood</td>
<td>10 mL</td>
<td>3 weeks</td>
</tr>
<tr>
<td>3</td>
<td>12.1</td>
<td>30.0</td>
<td>2 hours</td>
<td>Peri-orbital</td>
<td>No</td>
<td>No</td>
<td>Blood + furosemide</td>
<td>Blood</td>
<td>10 mL</td>
<td>5 days</td>
</tr>
<tr>
<td>4</td>
<td>7.8</td>
<td>30.0</td>
<td>34 hours</td>
<td>Occipital</td>
<td>Yes</td>
<td>Yes</td>
<td>Blood</td>
<td>Blood</td>
<td>18 mL</td>
<td>4 weeks</td>
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<tr>
<td>5</td>
<td>1.0</td>
<td>&gt;55.0</td>
<td>2 hours</td>
<td>Holocapthic</td>
<td>Yes</td>
<td>Yes</td>
<td>Blood</td>
<td>Blood</td>
<td>6 mL</td>
<td>3 weeks</td>
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<tr>
<td>6</td>
<td>15.0</td>
<td>23.0</td>
<td>7 days</td>
<td>Occipital</td>
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<td>Yes</td>
<td>Blood</td>
<td>Blood</td>
<td>3 mL</td>
<td>2 ½ years</td>
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<tr>
<td>7</td>
<td>10.8</td>
<td>24.2</td>
<td>34 hours</td>
<td>Occipital</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Blood</td>
<td>18 mL</td>
<td>4 weeks</td>
</tr>
<tr>
<td>8</td>
<td>7.0</td>
<td>27.0</td>
<td>48 hours</td>
<td>Frontal</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Blood</td>
<td>20 mL</td>
<td>6 weeks</td>
</tr>
<tr>
<td>9</td>
<td>8.0</td>
<td>22.0</td>
<td>12 months</td>
<td>Occipital</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Blood</td>
<td>42 mL</td>
<td>6 months</td>
</tr>
</tbody>
</table>

Note: N/A indicates data not available.

* Nausea/vomiting
* Received follow-up care at another institution.

RIH: Why does it happen?

We don't know for sure

Possible mechanisms:
- “Squeeze” on the sac, displaces CSF
- Compensatory ↑ CSF production
- Reduced compliance – venous dilation

RIH: What is the prevalence?

30 consecutive patients
Day +1 after epidural blood patch

Graded RIH:
- 0 = none
- 1 = mild
- 2 = moderate
- 3 = severe

RIH, severity:
- None 33%
- Mild 43%
- Moderate 17%
- Severe 7%
RIH: How to treat

- **Mild**
  - Elevate head
  - Analgesia

- **Moderate**
  - + Acetazolamide oral
  - +/- immediate acetazolamide IV

- **Severe**
  - LP to remove fluid

Other meds: (?)
- topiramate
- furosemide/HCTZ
- nortryptiline

**Rebound Intracranial Hypertension**

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