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The Varied Clinical Presentations of SIH

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Disclosures (past 2 years):

Role	Organization
Advisory Board	Allergan, Amgen, Avanir, Biohaven Pharmaceuticals, electroCore, Eli Lilly, Impel, Lundbeck, Satsuma, Teva, Zosano
Consultant	Avanir, Eli Lilly, electroCore
Support: Clinical trial site PI	Allergan, Eli Lilly, Zosano
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Medical Advisor	**Spinal CSF Leak Foundation,** HealthyWomen
Editorial Board	Neurology Reviews
Contributing author	Medlink Neurology, Medscape

Bold = relevant to content

Case 1

- 31-year-old woman with diplopia and intermittent headaches that became constant in 2 weeks
- Optometrist → right abducens palsy → normal CT orbits
- Evaluated 2 months later
  - Right temple pain with intermittent burning of right cheek & ear
  - Sharp retro-orbital pain and pain to right of vertex
  - Dull pain in right neck and occiput
  - Severe photophobia, mild phonophobia, nausea, confusion, dry heaves
- Pain on awakening 4 out of 10; 8 out of 10 at the end of the day
- Worse with coughing, sneezing, bearing down, standing
- Improved lying flat, caffeine
- Normal brain MRI with contrast

Case 2

39-year-old music teacher with migraines with visual aura since age 30, occurred with menses and one other time per month, relieved with rizatriptan.

May 3, 2014 – Went on a band trip and developed migraine lasting 6 weeks; not healthy since then

Rizatriptan was ineffective, steroids helped for one day

Started a migraine diet

Fluorescent lights in school were bothersome

Photophobia gradually worsened

- September 2014 – could drive, tolerate sunlight and fluorescent lights at school
- Spring 2015 – could tolerate incandescent light
- Summer 2015 – could eat dinner by candlelight
- August 2017 – has to eat in the dark

At time of visit: All light exposure triggers a migraine

Extremely photophobic with scalp allodynia

Current Headaches

Feel like a knife stabbing in her head, or a skewer in her head, or like acid burning

Bilateral head pain with neck pain at times

Photophobia, phonophobia, osmophobia

No nausea unless she rides in the car

Trouble concentrating with a severe headache

Off balance, as though she will fall

If she starts vomiting, she has to go to the ER because home medications don’t work. Sometimes also gets diarrhea.

Examination

Normal neuro-ophthalmic exam

Affect appropriate

Wore sunglasses in the office, covered her head with a towel when possible (especially when exam room door opened)

Covered computer monitor with a towel when not in use

Brought her own floor lamp to the exam (38 watt incandescent bulb)

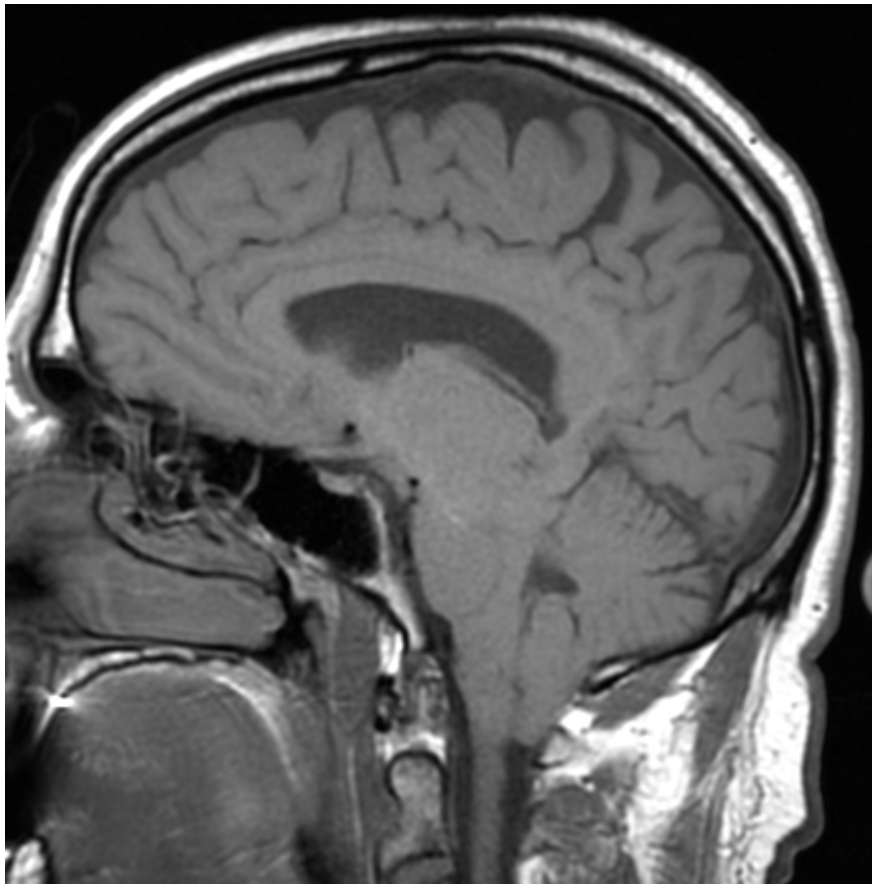
MRI brain with contrast (February 2014) reviewed, normal

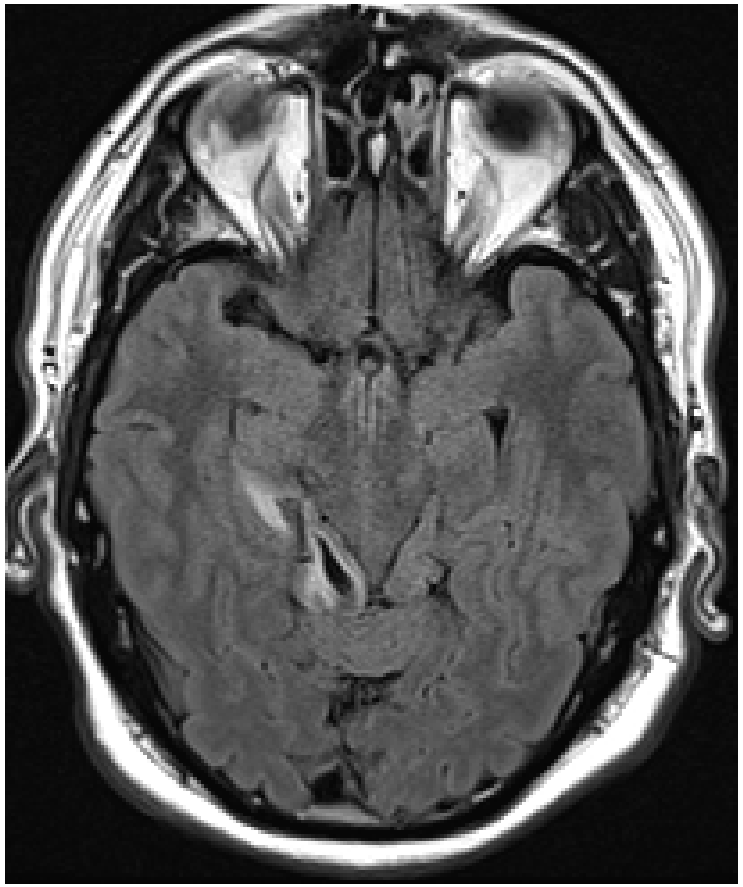
Case 3

61-year-old director of treasury for large company
3 year history of cognitive decline
- Missed paying bills
- Slept at his desk, couldn't log into computer
- Could not recall the day of the week

Headaches started 2 years later, 2-3 times weekly
- Top & back of head, neck; photophobia only
- Last 30-60 minutes, relieved with acetaminophen
- No postural component or change with Valsalva

No imbalance, tinnitus, hearing problems. Walks more slowly.
Referred after memory clinic obtained MRI
Wife related his history in the office





Case 4

65-year-old retired professor of nursing

Persistent headache following a spinal tap in 2014

- Worse with any positional change
- Bifrontal, aching, vomiting when severe

6 months later, started shaking and developed generalized weakness (using a walker)

Diagnosed with meningitis when MR showed dural enhancement

Memory problems (better in the AM), off balance, falls → living with her mother

Case 5

- 46-year-old woman with orthostatic headaches for 10 years
- Occurred after being upright for 6-7 hours, 7 out of 10
- Top of head, sharp with nuchal aching
- Photophobia, constant tinnitus; pulsatile tinnitus in the AM
- Daily, constant
- Relieved only with sleep and at high altitude
- Also with occipital headaches and interscapular tension and burning neck pain

1 year prior she woke up two days in a row with a “wet ear” and a halo of blood and clear liquid on the pillowcase

Headaches worsened after this

Evaluation for skull base CSF leak was negative

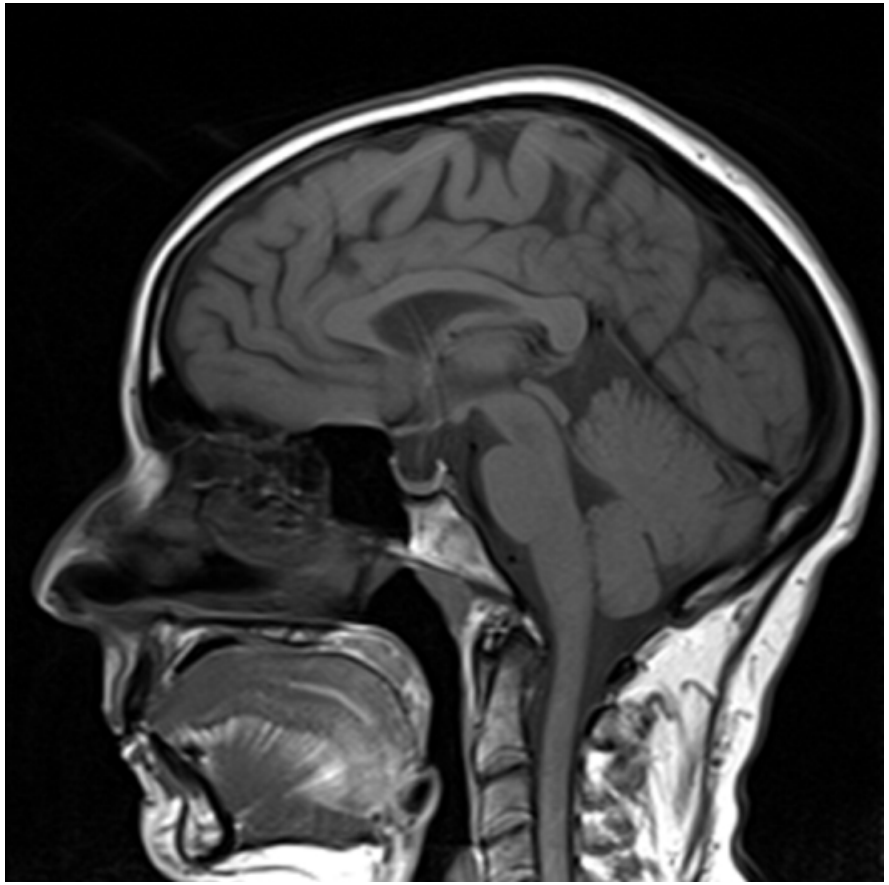
Started on topiramate 100 mg daily

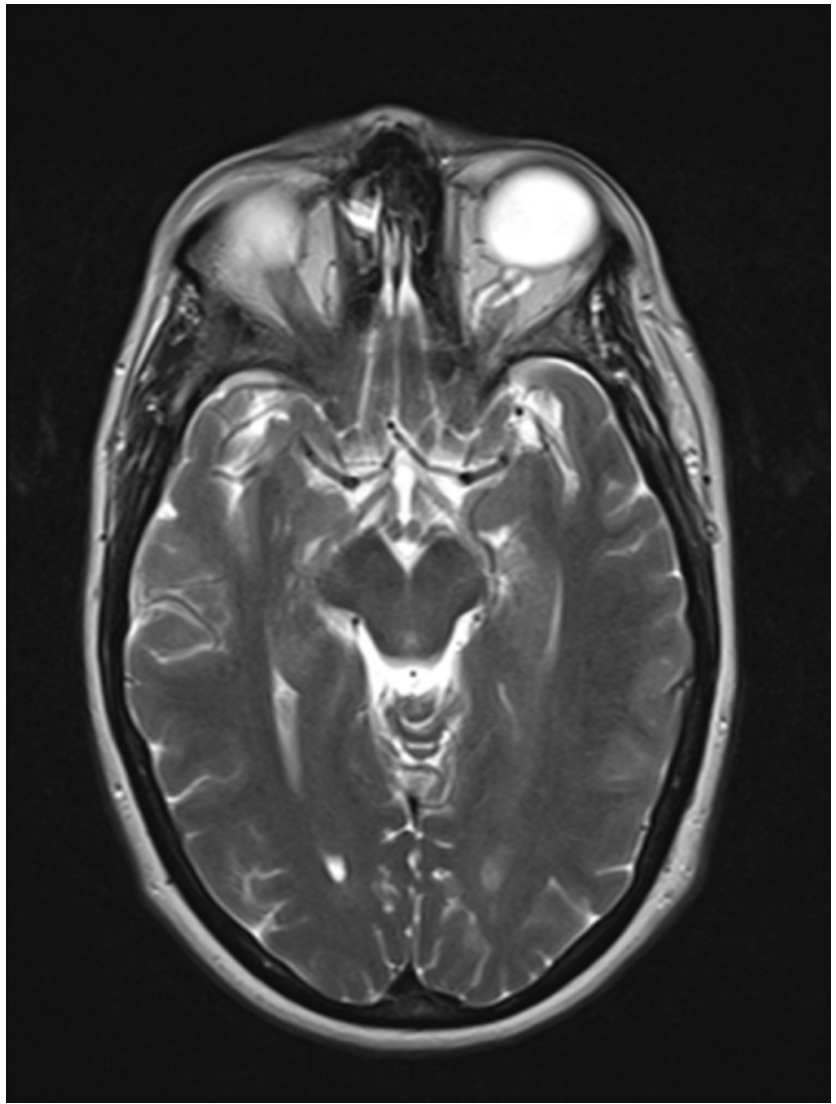
LP 5 years prior for possible IIH showed OP 150 mm CSF

CT myelogram showed multiple perineural cysts but no leak

Headaches improved for 1 month after non-targeted blood patch

Imaging 2 years prior





- PMHx: Ehlers Danlos syndrome

- Exam: BMI 28 kg/m^2

  Normal optic nerves with spontaneous venous pulsations

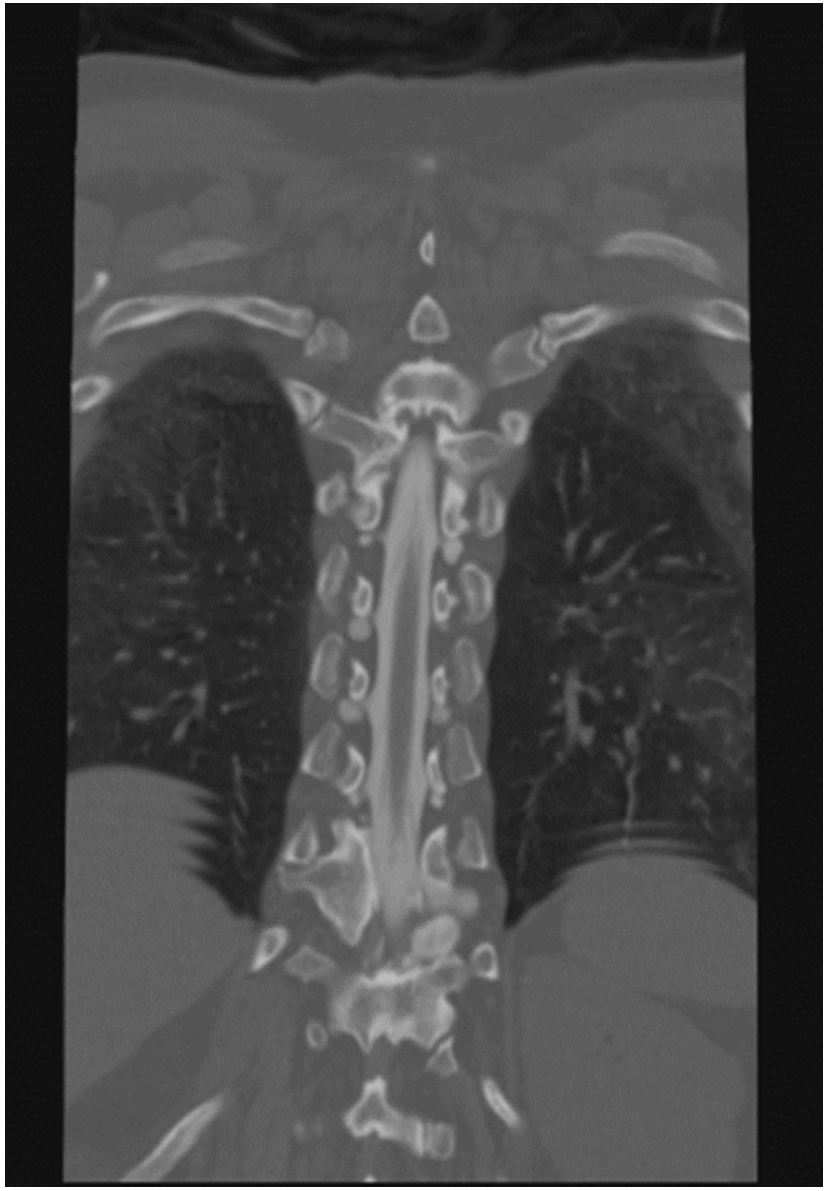
  Normal neuro exam

  Trendelenburg test: 7→ 5 out of 10 in 10 minutes

Targeted blood patch of perineural cyst at T10-11 gave short lived relief

Topiramate discontinued for possible exacerbation of intracranial hypotension

Subsequent blood patches with relief for 5- 9 weeks



Developed a different headache10 days after her last blood patch

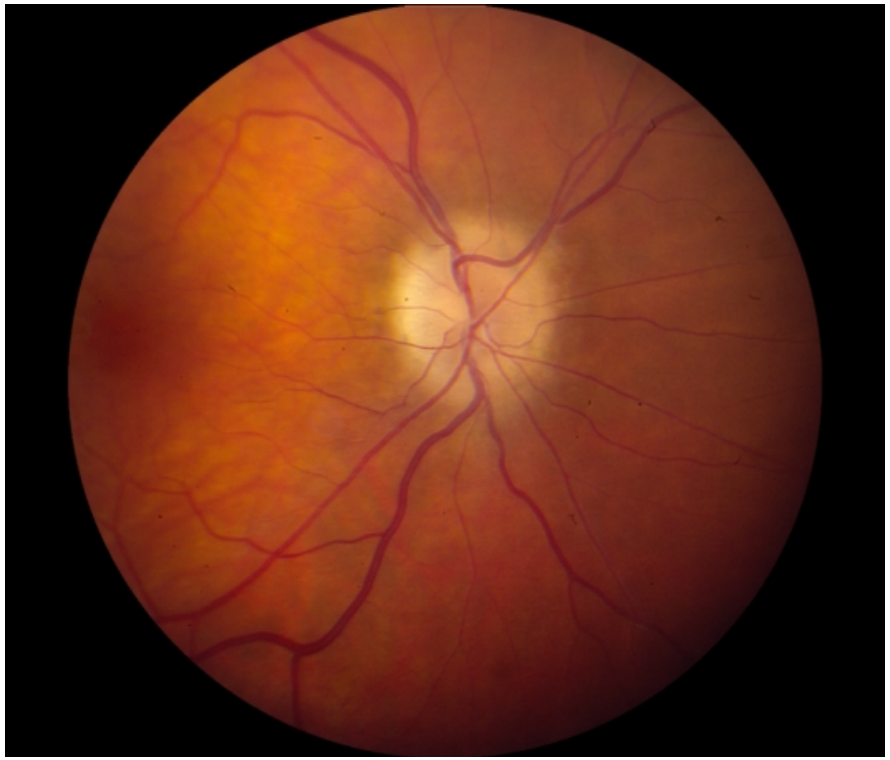
Worse when lying flat

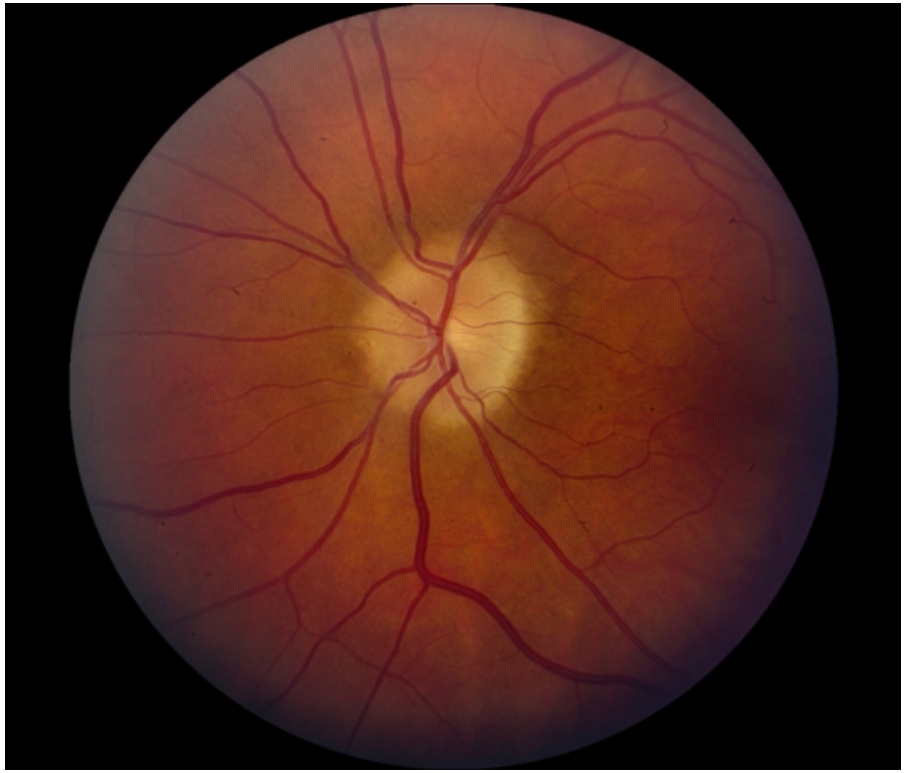
Awakened with headache that resolved 10-15 minutes of being upright, then the previous orthostatic headache began 4 hours later

More history….

Gained 30 pounds after stopping topiramate

“Life long” history of transient visual obscurations when standing





Case 6

- 51-year-old nurse with previous history of migraine without aura controlled with topiramate and sumatriptan
- T-boned in a car accident April 2019. Airbag deployed. Sustained brief LOC, jaw fracture, neck pain, rib contusions
- 2 months later had thunderclap headache during sexual activity (orgasm), located posteriorly. Sharp, shooting pain lasting 6 hours “thought I was going to die”.
- Continues to have severe headaches only while approaching orgasm, heavy lifting, straining
- Also has cognitive problems (medical coding class)

Seeing a Headache Specialist for

- Severe photophobia
- Dementia, Cognitive decline
- “Functional” Tremor
- Abducens Palsy
- Burning Neck Pain
- Facial Pain

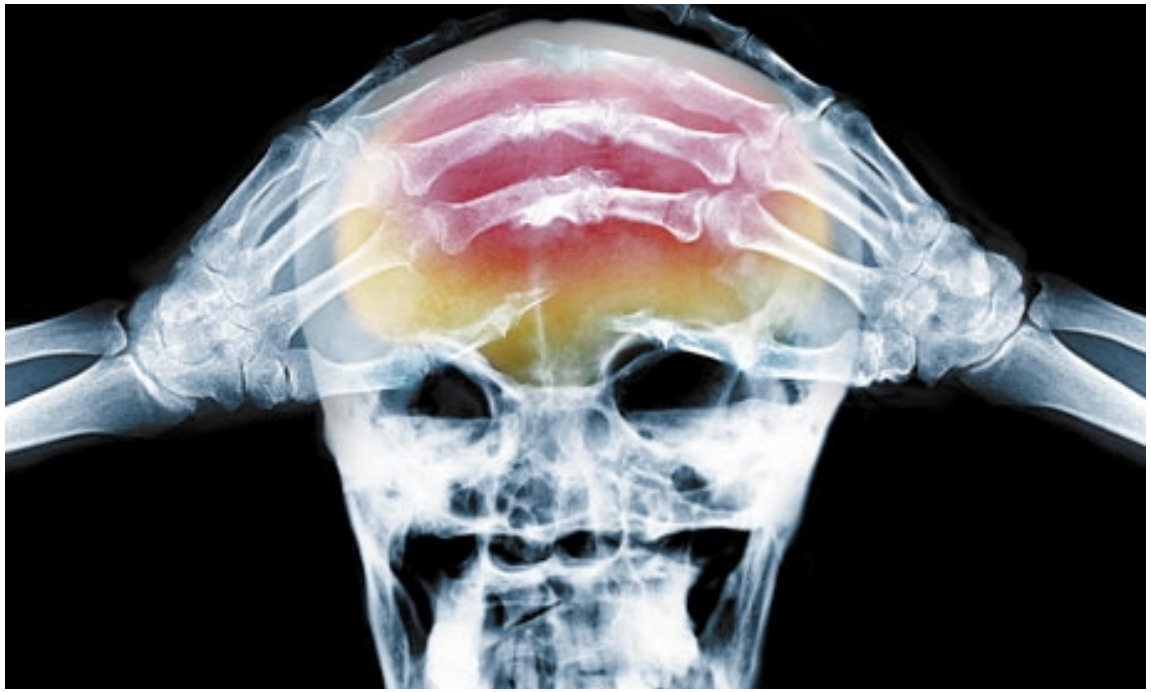
These patients are in your practice!

They are not necessarily the same patients who see neurosurgeons and neuroradiologists (referral bias)

Only 70% of patients are “typical”

You need to be a detective.

Who Sees a Headache Specialist?



Headache in SIH

Not always present, even with abnormal MRI

New daily persistent headache

May be thunderclap in onset

Most commonly posterior/neck but may be in any location in the head (and face)

Pain is most frequently bilateral
No specific character or severity

Typical: Orthostatic or “end of the day”

May be paradoxical (better when upright)

Postural component may decrease over time

May be intermittent or constant

Nocturnal awakening possible

Often exertional and worsens with Valsalva

May improve at high altitude

Other symptoms

- Photophobia, phonophobia, nausea, vomiting
- Tinnitus, abnormal hearing (under water), imbalance (VIII)
- Spinal symptoms: neck pain, interscapular pain (“coat hanger headache”), chest pain, radicular symptoms, pain at the site of the leak
- Hypo and hyperkinetic movement disorders
- Cognitive dysfunction
- Coma
- Subdural fluid collections
- Intracranial hemorrhage
- Galactorrhea
- “Any cranial nerve”

Any Cranial Nerve?

CN	Possible Symptoms and Signs	Frequency
I	Altered taste, hyposmia, phantosmia, osmophobia (HA?)	Uncommon
II	Photophobia (HA?) Blurred vision Visual field defects, transient obscurations of vision Normal-tension glaucoma	Very common Uncommon Rare Rare
III	Transient or incomplete palsy	Rare
IV	No reports	No reports
V	Headache, facial pain, jaw pain, numbness	Very common
VI	Horizontal diplopia	Uncommon (most iatrogenic or secondary cause)

References listed at the end

CN	Possible Symptoms and Signs	Frequency
VII	Facial palsy, hemifacial spasm, facial grimacing	Uncommon
VIII	Hearing loss, muffled hearing (underwater), aural fullness Tinnitus Benign Paroxysmal Positional Vertigo (BPPV) Nystagmus Imbalance	Common Common Uncommon Rare Common
IX X XII XII	Vocal cord paralysis Orthostatic dysphonia, hoarseness, cough Dyspnea, dysphagia Throat discomfort Torticollis (focal dystonia) Dysgeusia	Rare Rare Rare Rare Rare Rare

References listed at the end

Epidemiology

Female: Male = 2:1

Peak incidence around age 40 but can occur at any time of life

Annual incidence at least 5/100,000

(*no ICD code until 2020)

May go undiagnosed for many years

Risk Factors

- Joint hypermobility syndromes (e.g., Ehlers Danlos, Marfan, polycystic kidney disease)
- Trauma – may be trivial
- Previous spine surgery
- Discogenic microspurs (ventral leaks)
- Previous LP, epidural or spinal anesthesia
- History of spontaneous retinal detachment

Beck J et al. Neurology 2016;87:1-7
Schievink WI. JAMA 2006;295:2286-96

When To Suspect SIH

- New daily persistent headache
- Chronic migraine, refractory to “everything”
- Patients with joint hypermobility
- Daily headache, lives in the dark
- Headache provoked by exertion, Valsalva maneuver

Questions to Ask

Headache:

- How do you feel when you first wake up, before you get out of bed?
- Is the headache better or worse at a particular time of the day?
- Effect of coughing, sneezing, straining, lifting, bending, exertion, laughing, singing, talking, sexual activity?
- Nocturnal awakening?
- Effect of caffeine (The F Word)?
- If better with recumbence, how long does it take to improve(not to be confused with sleep)?
- Effect of high altitude (plane, travel)?

Joint Hypermobility

"Double jointed", super flexible?

Good at yoga (shoulder wrap, touch the floor with straight legs)?

Flexible as a child? (splits, wrap legs around head)?

Active in sports: Gymnastics, ballet, tumbling, cheerleading

Family history of aneurysm, dissection, retinal detachment at a young age, non-rheumatic valvular heart disease?

(Can use validated scales on exam)

Neilson D, Martin VT. Headache 2014;54:1393-402
Martin VT, Neilson D. Headache 2014;54;1403-11

Precipitating Events and Other Risk Factors

- Injury (MVA, fall)
- Chiropractic manipulation
- Roller coaster
- Violent coughing
- Protracted vomiting
- “Epidural” anesthesia or block
- LP, myelogram, spinal anesthesia
- Heavy lifting (gym, moving)

- Athletic activities, especially involving twisting: golf, tennis, canoeing, kayaking, yoga, fly fishing
- Working out/lifting weights
- Previous spine surgery
- Known herniated or calcified disks
- Bariatric surgery
- History of “Chiari” or POTS
- History of EDS

Examination Findings

May have joint hypermobility

Reassuring if spontaneous venous pulsations present

Trendelenburg test (5° for 5-10 minutes)

- May improve headache and other symptoms

Say…are you “double jointed”?

Any Cranial Nerve References

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