Atypical Presentations of Spinal CSF Leaks

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Disclosures (past year):

Role	Organization
Advisory Board/Consultant	AbbVie, Amneal, Axsome, Scilex, Tonix
Speakers Bureau	AbbVie, Scilex
Medical Advisory Board	Healthy Women, **Spinal CSF Leak Foundation,** Migraine Meanderings
Editorial Board	Neurology Reviews, J Neuroophthalmology
Contributing author	Medlink Neurology,

Bold = relevant to content

Case 1

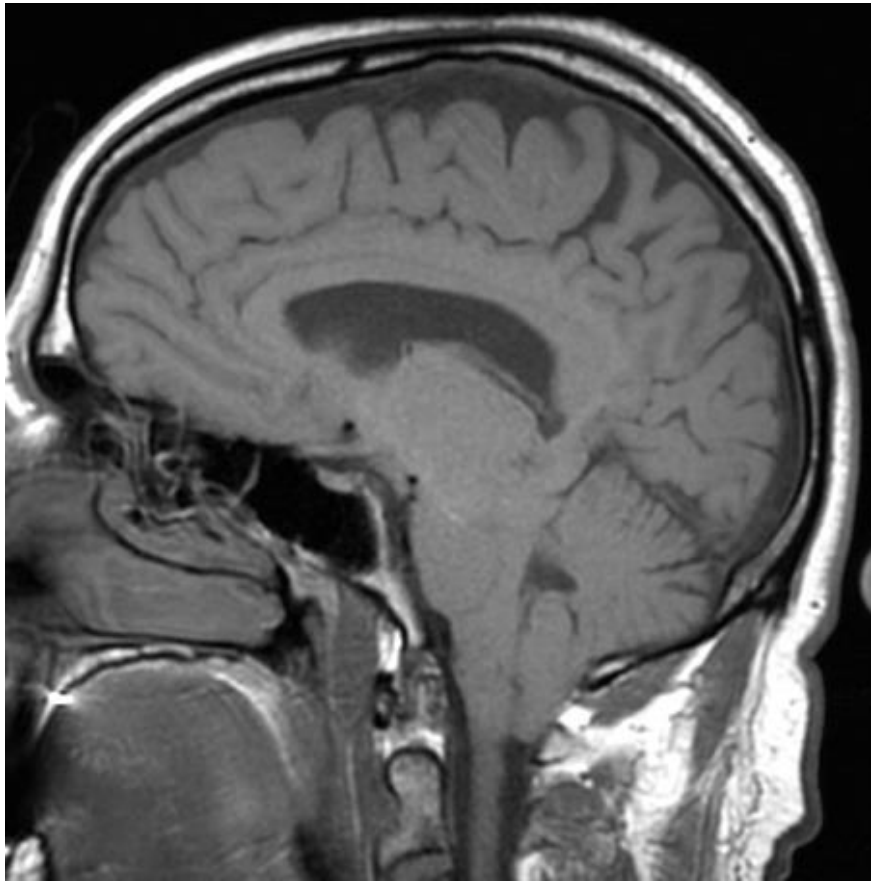
- 31-year-old woman with diplopia and intermittent headaches that became constant in 2 weeks
- Optometrist → right abducens palsy → normal CT orbits
- Evaluated 2 months later
  - Right temple pain with intermittent burning of right cheek & ear
  - Sharp retro-orbital pain and pain to right of vertex
  - Dull pain in right neck and occiput
  - Severe photophobia, mild phonophobia, confusion, nausea,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

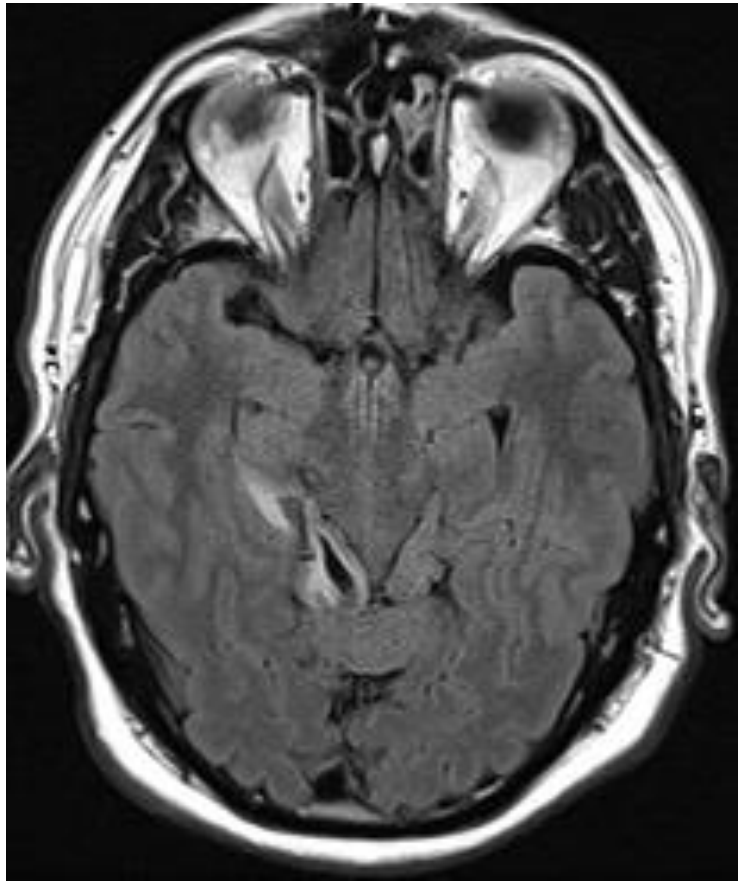
Case 2

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 3

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 4

64-year-old woman with rapid onset of orthostatic headaches 4 days after having a skull injury when a heavy screwdriver fell on her head from ~4 m in height

Rapid onset of orthostatic headache

Bilateral upper-lid ptosis

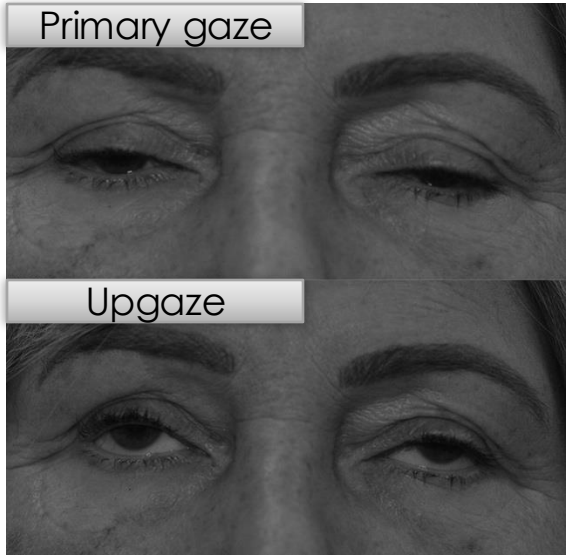
Blurred vision but no diplopia

Previous eye exams normal

MRI showed bilateral subdural fluid collections, midbrain sag, cerebellar tonsillar descent, loss of the ambient cistern

Vahdani K et al. Orbit 2018;37(5):371-4

- Exam showed normal acuity, bilateral ptosis and generalized external ophthalmoplegia, hypometric saccades
- Headaches resolved after 3 EBPs – no change in eyes
- Had bilateral brow suspension at 18 months
- Eye movements unchanged but no diplopia



Vahdani K et al. Orbit 2018;37(5):371-4

Case 5

53-year-old man with 7-year history of slowly progressive, asymmetric brachial amyotrophy, left>right.

2008 - EMG suggested pathology of cervical motor roots or anterior horn cells. Diagnosed with atypical motor neuron disease ("ALS")

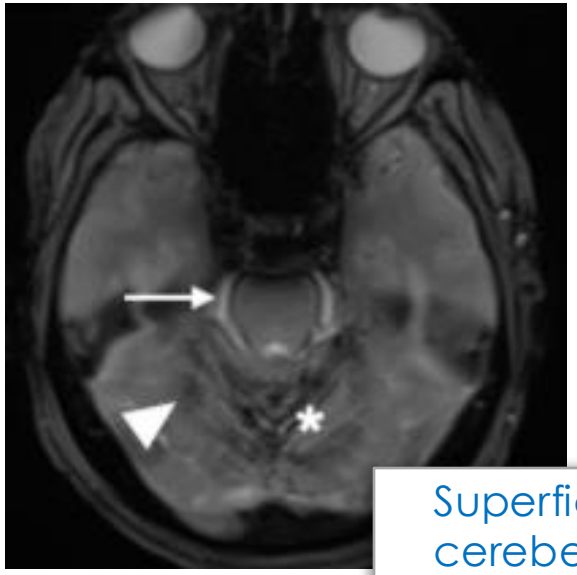
2019 – worsening upper limb weakness and wasting with sensorineural hearing loss

MRI showed extensive supra- and infratentorial superficial siderosis (surface of entire spinal cord), and large ventral intraspinal fluid collection with bony spurs at C6-C7

2021- developed parkinsonism, responded to levodopa (felt to be idiopathic PD and unrelated)

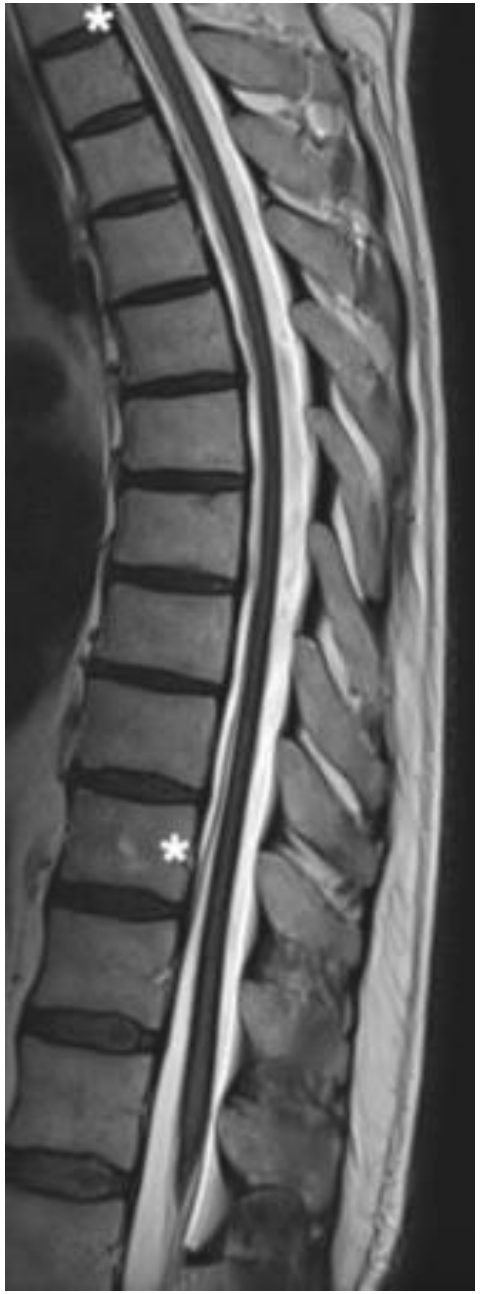
2022 -no change in symptoms, exam or imaging

Iakoleva V et al. BMC Neurol 2024;24:309



Superficial siderosis of cerebellum and pons (GRE image)

ALS-like picture attributed to the dural leak with compression of cervical motor roots.
Superficial siderosis was secondary to the dural tear and chronic bleeding.



Hypointense rim of hemosiderin along the length of the spinal cord with extradural fluid collection from T2-T11 (*)

Iakoleva V et al. BMC Neurol 2024;24:309

Case 6

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 – had 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

Severe photophobia

"Functional" Tremor

Chronic progressive external ophthalmoplegia (CPEO)-like picture

Dementia

Abducens Palsy

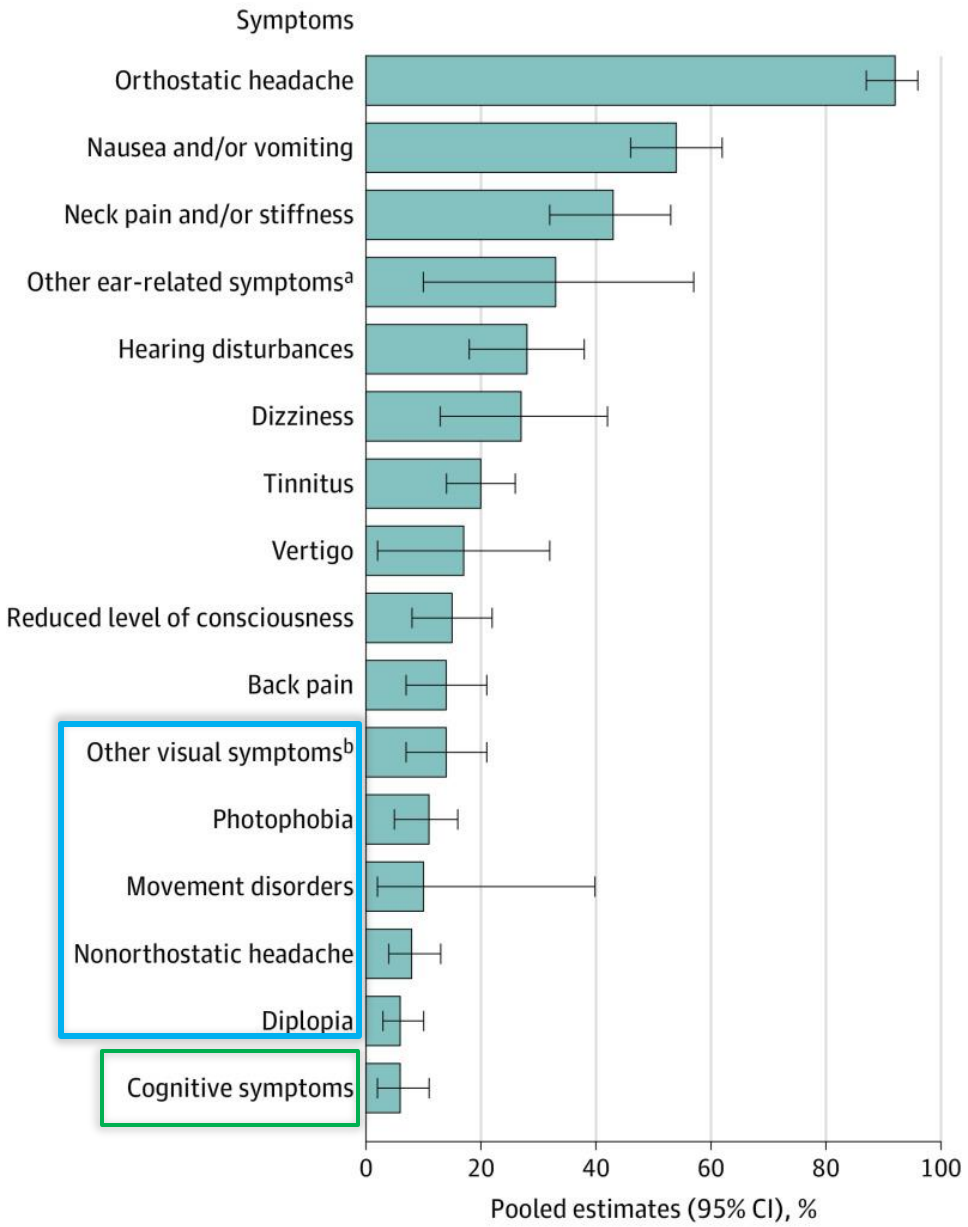
"Burning acid" on the head

ALS-Like picture with secondary superficial siderosis

Clinical Presentation – A Meta-Analysis

- Method: PRISMA reporting guideline-compliant systemic review and meta-analysis of SIH literature through April 30, 2020
- Sources: PubMed/MEDLINE, Embase, Cochrane
- Study selection: English language reporting 10 or more patients
- Outcome: Pooled estimate proportions of SIH symptoms, imaging finding and treatment outcomes
- Results: 144/6878 articles met criteria with 53 patients on average (range 10-568)

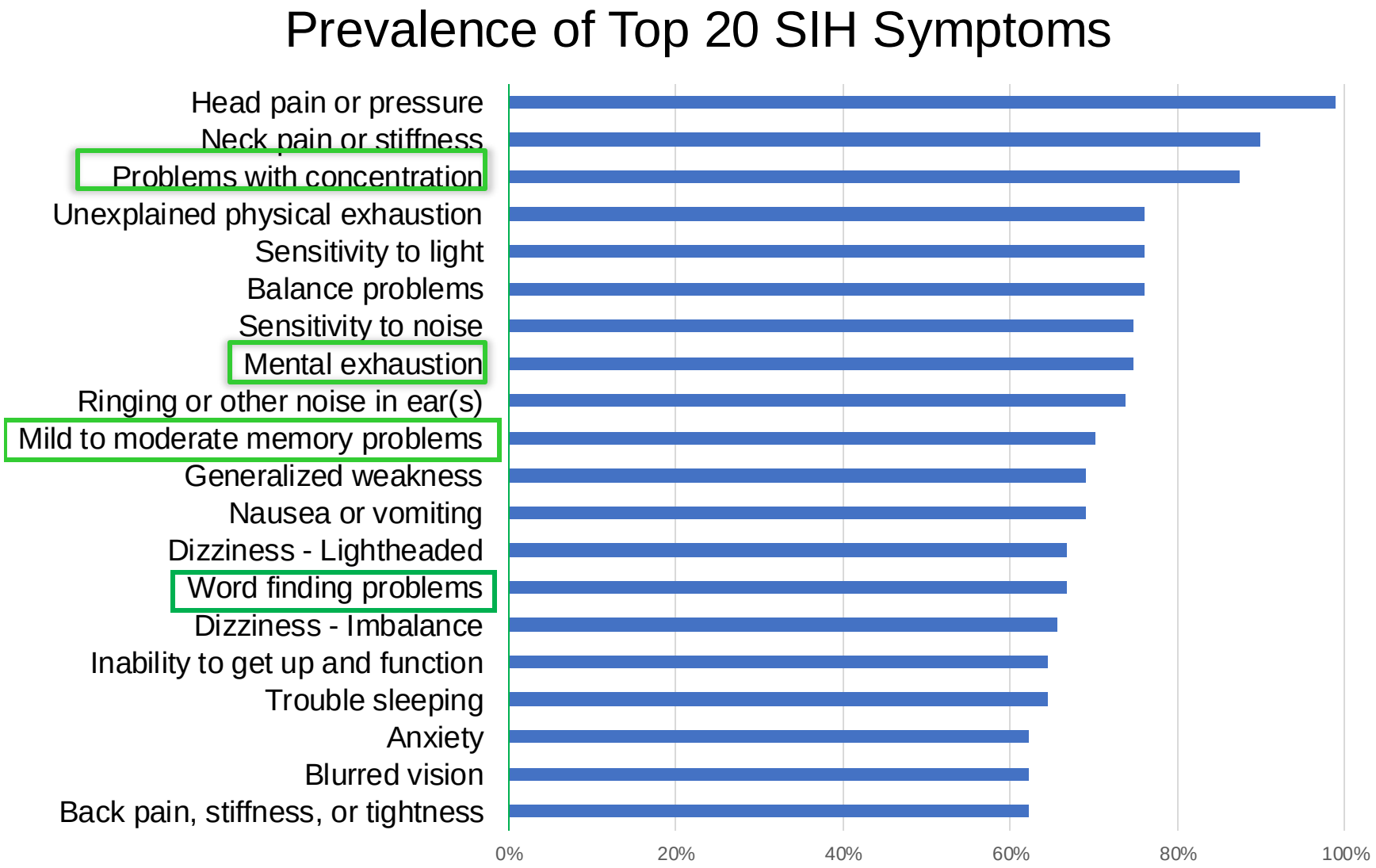
D'Antona A et al. JAMA Neurol 2021;78:329-337



- Mean age 42.5 years (range 2-88)
- 63% females
- Risk factors: connective tissue disorders, spinal pathologies (i.e., osteophytes, disc prolapse, discogenic microspurs)
- Headache was most commonly diffuse, occipital or frontal
- 2.3% had non-orthostatic headache
- 1.4% had no headache

D'Antona A et al. JAMA Neurol 2021;78:329-337

UT Southwestern Experience – SIH Impact Inventory (n=98)



62 (63.3%)confirmed SIH
36 (36.7%) suspected SIH

Friedman DI et al. American Headache Society Annual Scientific Meeting 2024

Other Unusual Manifestations

- Visual field defects (binasal)
- Oculomotor (III) nerve palsy
- Movement disorders (chorea, torticollis, parkinsonism)
- Spells of altered consciousness
- Gait ataxia upon arising
- Worsening (or improving)at high altitude
- Galactorrhea
- Speech and swallowing problems
- Dysautonomia and orthostatic intolerance

Horton JC, Fishman RA. Ophthalmology 1994;101(2):244-51
D'Antona A et al. JAMA Neurol 2021;78:329-37
Nowak DA et al. J Neurosurg 2003;98(4)903-7
Batsis JA, Py MP. Headache 2005;45(4):380-4
Ghosh A et al. Child Neurol Open 2021;8:2329048X11056709

Conclusions

- While almost all patients have orthostatic headache, some may have minimal or no headache…and
- SIH can produce some very unusual symptoms, delaying or even "excluding" the diagnosis
- Cognitive impairment ("brain fog") and photophobia may be more common than previously recognized

Kim DK et al. Neurology 2021;96:e1312-18

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