31 y/o female with H/O positional headache, sensitivity of light, and nausea

Jignesh Modi, MD

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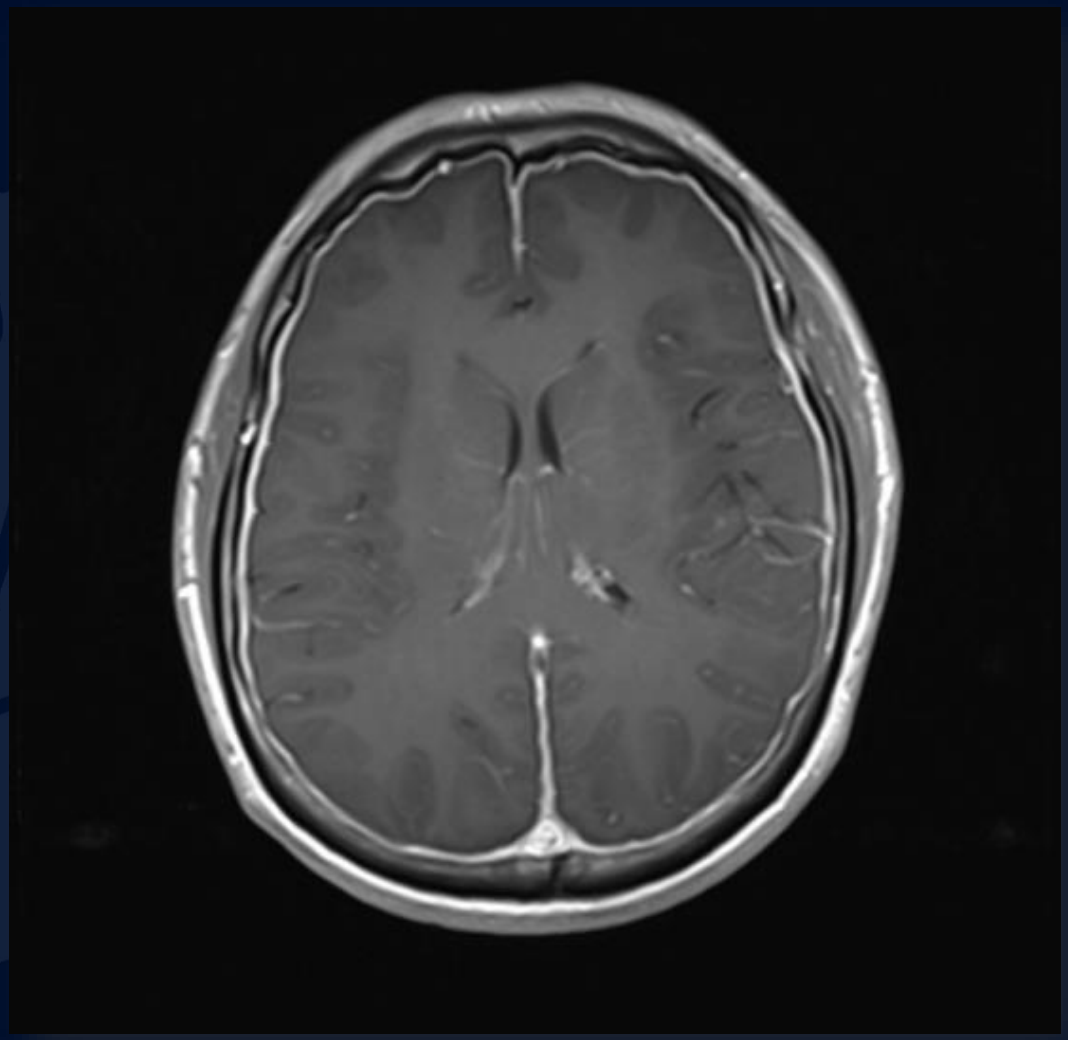

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Spontaneous Intracranial Hypotension

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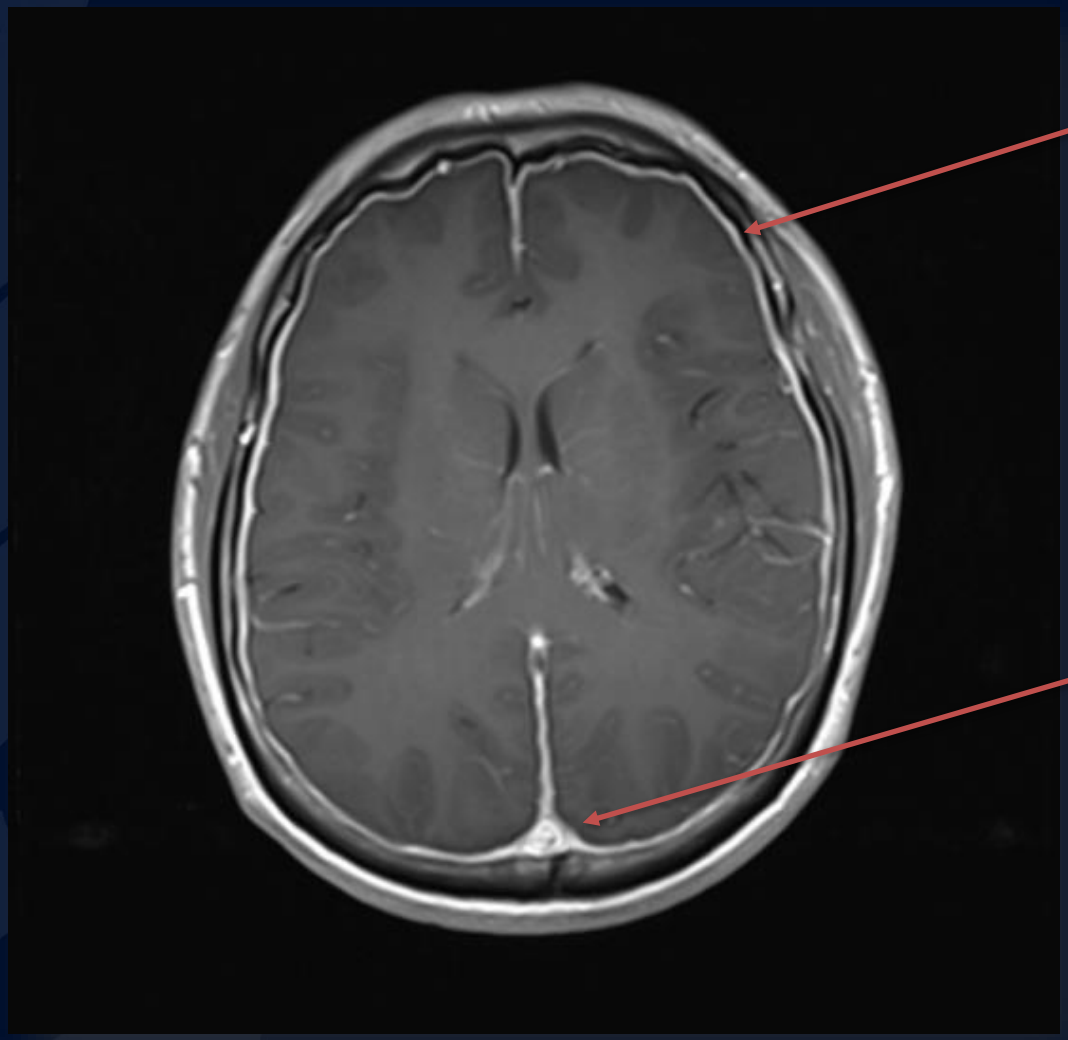

Diffuse pachymeningeal enhancement

Dural Venous Distension Sign

Axial T1 Post-contrast

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Sagittal T1 Pre-contrast

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Intracranial Hypotension

Primary – Spontaneous intracranial hypotension (SIH)

Secondary – Iatrogenic (lumbar puncture or surgery), over shunting due to diversion devices, or traumatic

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Spontaneous Intracranial Hypotension (SIH)

Pathophysiology: Low CSF volume secondary to leakage through a dural defect with no identifiable cause.

Epidemiology: 5/100,000 per year, female > male, prevalence is underestimated due to underdiagnosis.

Presentation

- Classic: Orthostatic headache (nonspecific and initial misdiagnosis is common)
- Other: Vague neurological symptoms such as non-positional daily headache, cranial nerve palsy, gait disturbance, and cognitive dysfunction.

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Imaging Appearance on Brain MR

- SEEPS
  - Mnemonic subdural fluid collection
  - Enhancement of meninges
  - Engorgement of venous structures
  - Pituitary Hyperemia
  - Sagging of the brain
- Qualitative Signs
  - Pachymeningeal (dural) enhancement
  - “Sagging of brainstem and low-lying cerebellar tonsils”, “interpeduncular angle”, “rule out other diagnoses”
  - Pituitary Hyperemia
  - Subdural effusions
- Quantitative Signs
  - Mamillopontine distance < 5.5 mm
  - Pontomesencephalic angle < 50 degrees
  - Interpendencular angle < 40.5 degrees
- Contrast-enhancement brain MR is first-line to confirm the diagnosis and rule out other diagnoses.

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CSF Leak Site

- Source of the leak remain occult in 46-55% patients despite advanced imaging
- Three main causes
  1. Leaking diverticulum
  2. Calcified disk
  3. CSF-Venous fistula

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Find Source of Leak

- CT myelogram
- Decubitus myelogram
- Digital subtraction myelogram
- MR myelogram, rarely

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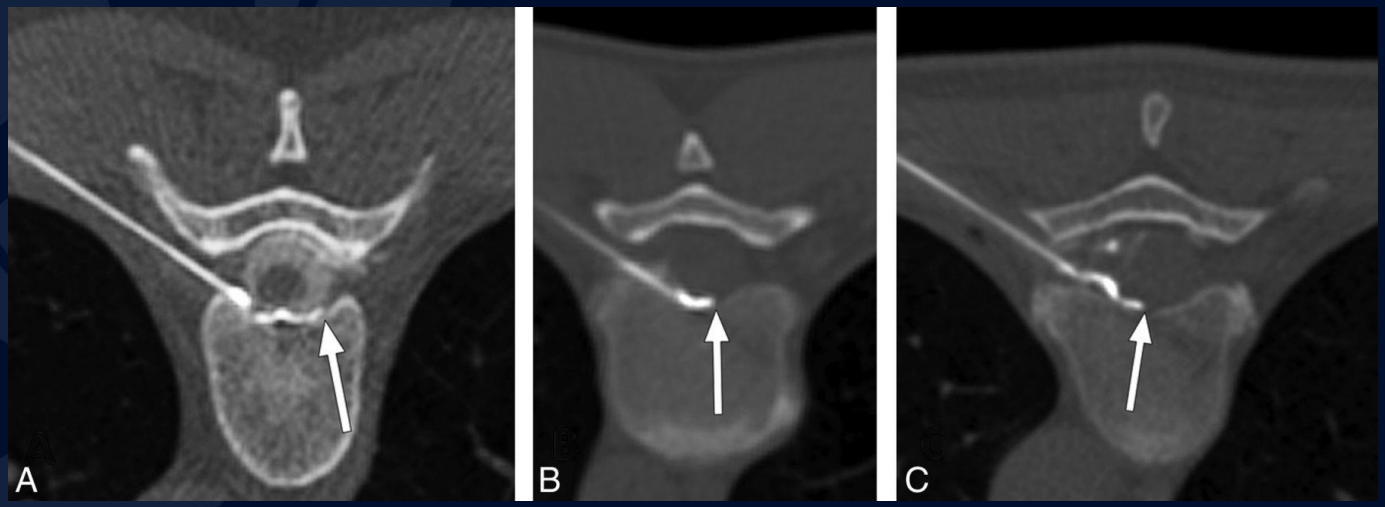
Treatment Approach

- Targeted vs. Non-targeted
- Blood patch
- Fibrin glue
- Surgery
  - Dural repair



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Post-Treatment

- Back pain for 24 hrs
- Rebound intracranial hypertension: Higher pressure, worse when laying down, often frontal, periorbital, vertex
- Follow up
  - Often need more than one patch
  - Follow up imaging

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Summary

- Familiarity with the spectrum of presentations and causes of SIH is critical for accurate and timely diagnosis and management.
- Remember the classic imaging appearance of brain sagging, diffuse dural enhancement for diagnosis with use of measurements in difficult cases.
- Treatment based on type of leak & cause.
- Challenges exist in both diagnosis and treatment. It requires understanding of the underlying pathogenesis of the condition. Prospective studies are needed for treatment decisions.

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