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

Jignesh Modi, MD

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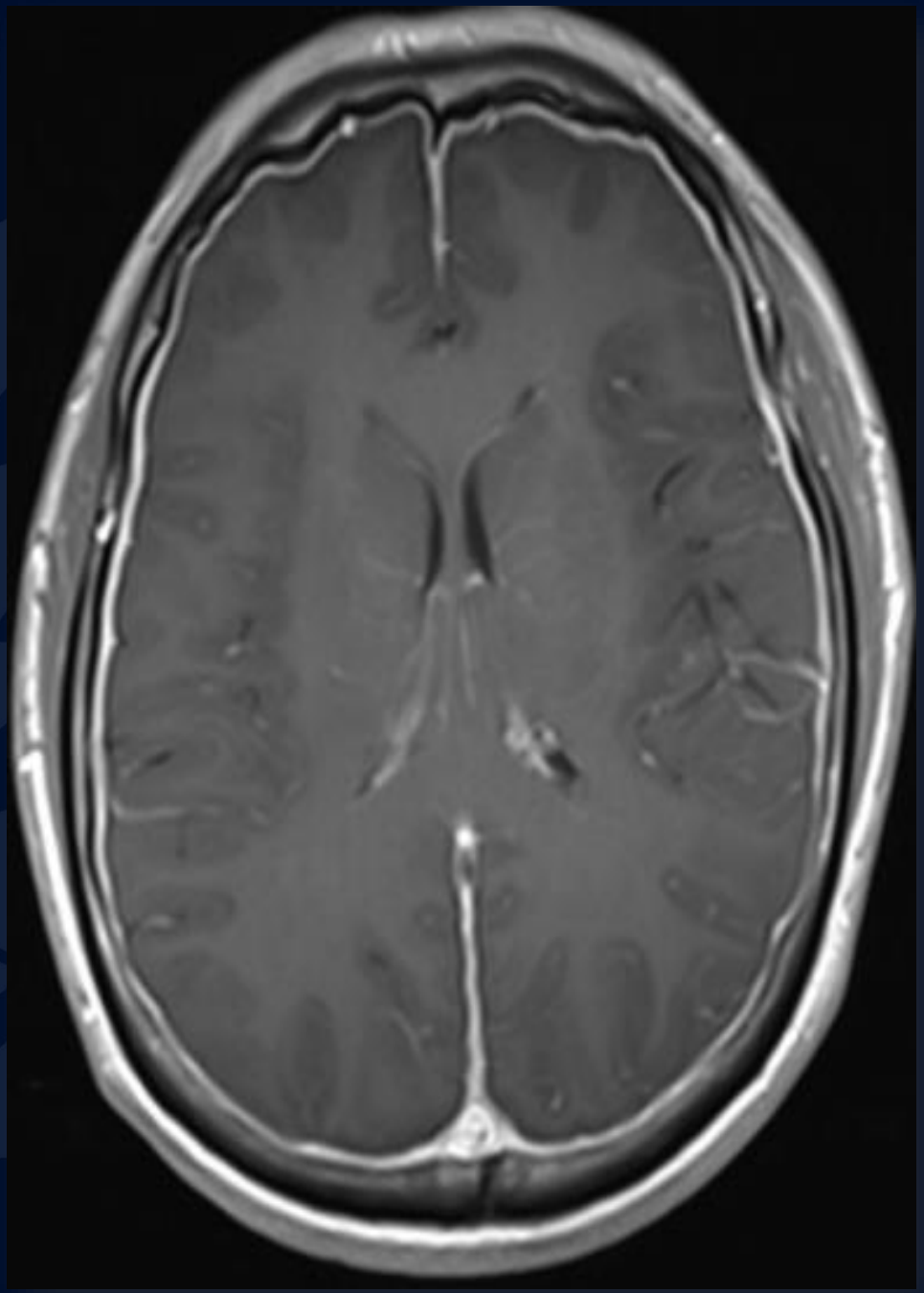

Axial T1
Post Contrast

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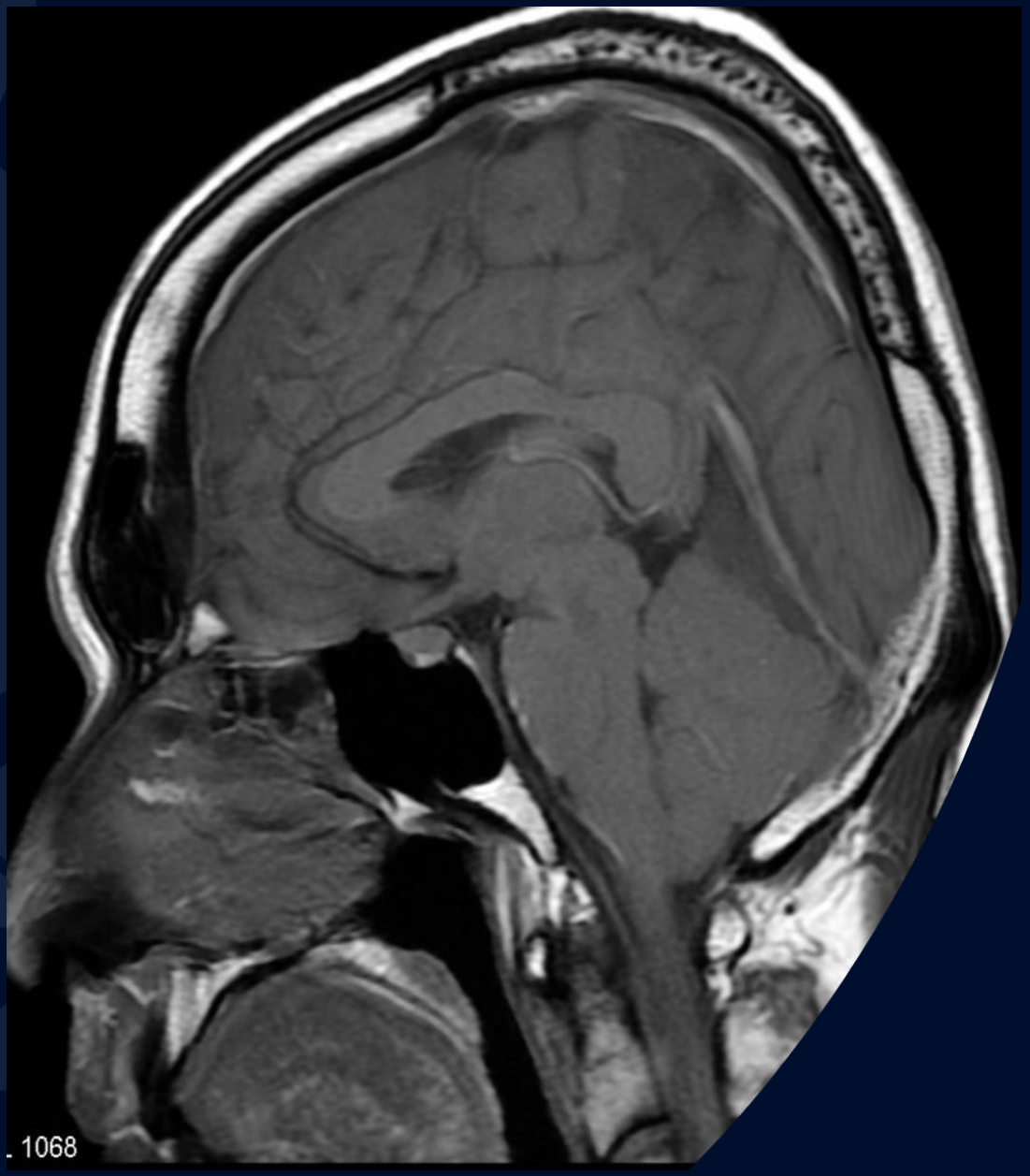

Sagittal T1 Pre-Contrast

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Spontaneous Intracranial Hypotension

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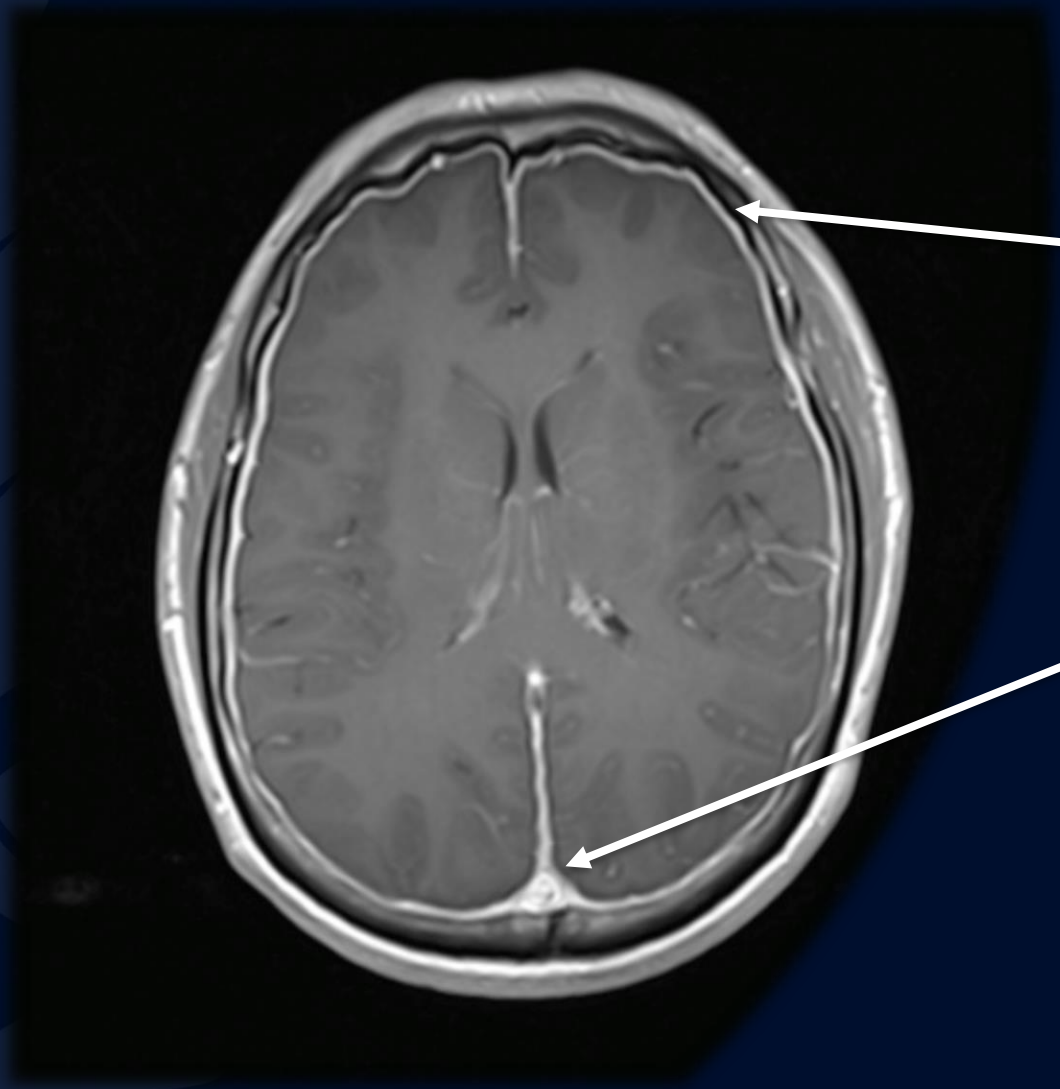

Diffuse pachymeningeal enhancement

Dural Venous Distension Sign

Axial T1
Post-Contrast

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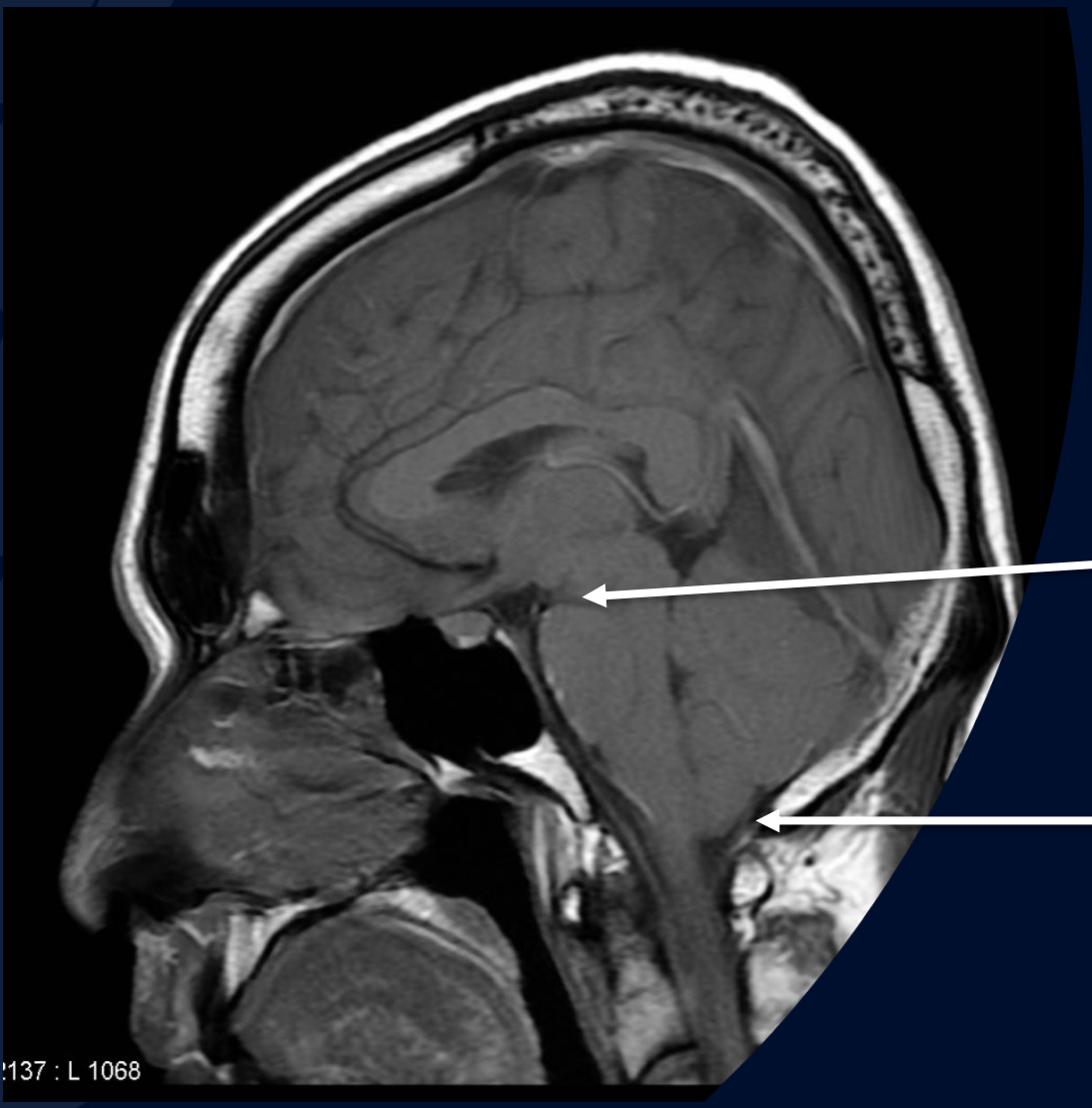

Sagittal T1
Pre-Contrast

Decreased mammillopontine distance

Low lying tonsils

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Intracranial Hypotension

1. Primary – Spontaneous intracranial hypotension (SIH)
2. Secondary – Iatrogenic (Lumbar puncture or surgery), over shunting due to diversion devices, or traumatic

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Spontaneous Intracranial Hypotension (SIH)

- Condition characterized by low CSF volume secondary to leakage through a dural defect with no identifiable cause.
- Incidence: 5/100,000 per year with majority of patients are female, however prevalence is underestimated due to underdiagnosis
- Classical presentation: Orthostatic headache (nonspecific and initial misdiagnosis is common)
- Many patients: Vague neurological symptoms: Non-positional daily headache, cranial nerve palsy, gait disturbance, and cognitive dysfunction.

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Imaging Appearance on MRI Brain

- 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

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Imaging Appearance on MRI Brain

- Quantitative Signs
  - Mamillopontine distance < 5.5 mm
  - Pontomesencephalic angle < 50 degrees
  - Interpendencular angle < 40.5 degrees
- Contrast-enhancement MR imaging of the Brain is the first-line investigation 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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Imaging modalities to find a leak for treatment approach

- CT MYELOGRAM
- DECUBITUS MYELOGRAM
- DIGITIAL SUBTRACTION MYELOGRAM
- Rarely: MR MYELOGRAM

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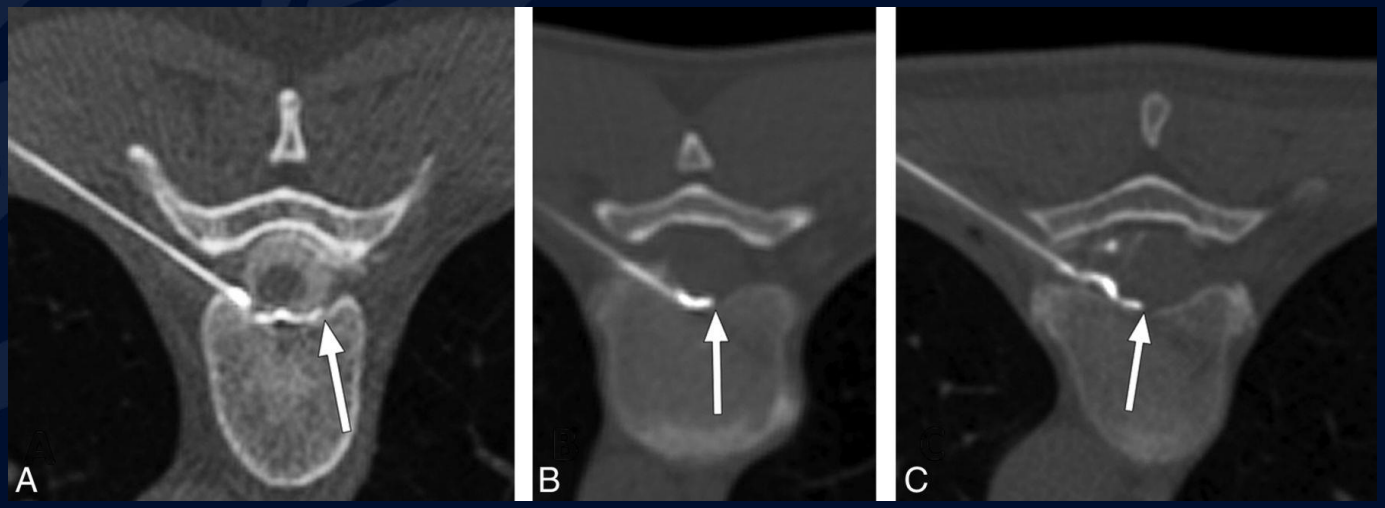
Treatment Approach

- 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:
  - "Usually 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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References

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