The story started with a phone call~

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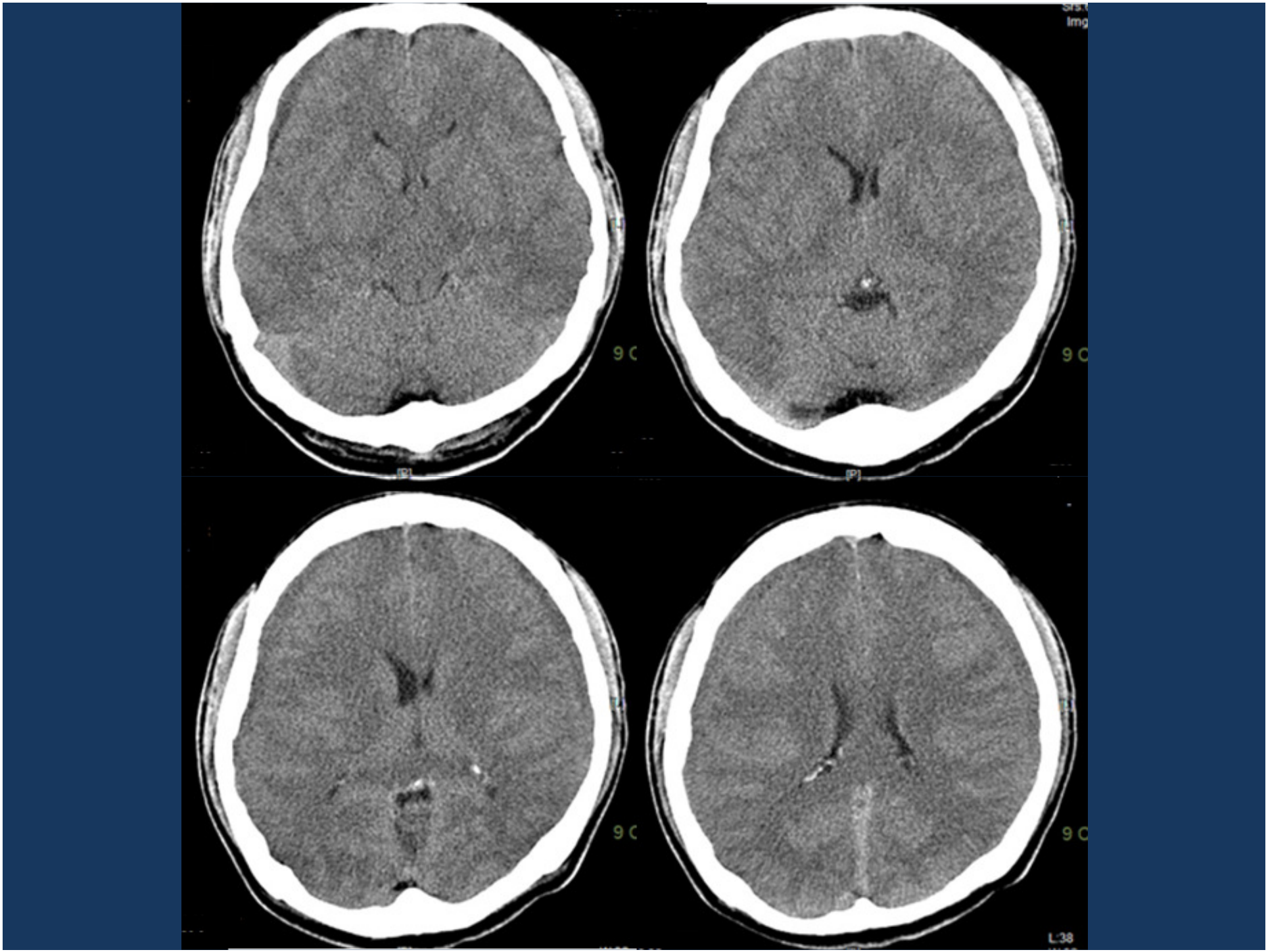


- 39 y/o police officer.
- One month ago, common cold with cough, sorethroat, rhinorrhea.
- Left temporal intermittent twitching pain. Pain score: 5/10. No blurred vision, nor other neurologic symptoms/signs.

- Cure from cold. Climbed mountain(higher than 3000m).
- Worsen diffused headache with mild neck stiffness, tinnitus.
- The headache happened about 10+ mins after standing from lying or sitting. Pain score: 10
- It was exacerbated with upright position and relieved with lying down.
- Disturbed his working and daily life.

Characters of headache

Location	Diffused, more severe over frontal area
Pain score	10/10
Quality	Dullness
Precipitating event	Upright position, especially 10+mins after standing up
Relieving factors	Lying down
Associated symptoms	Neck stiffness, tinnitus, neck pain with turning

Neurology Examination

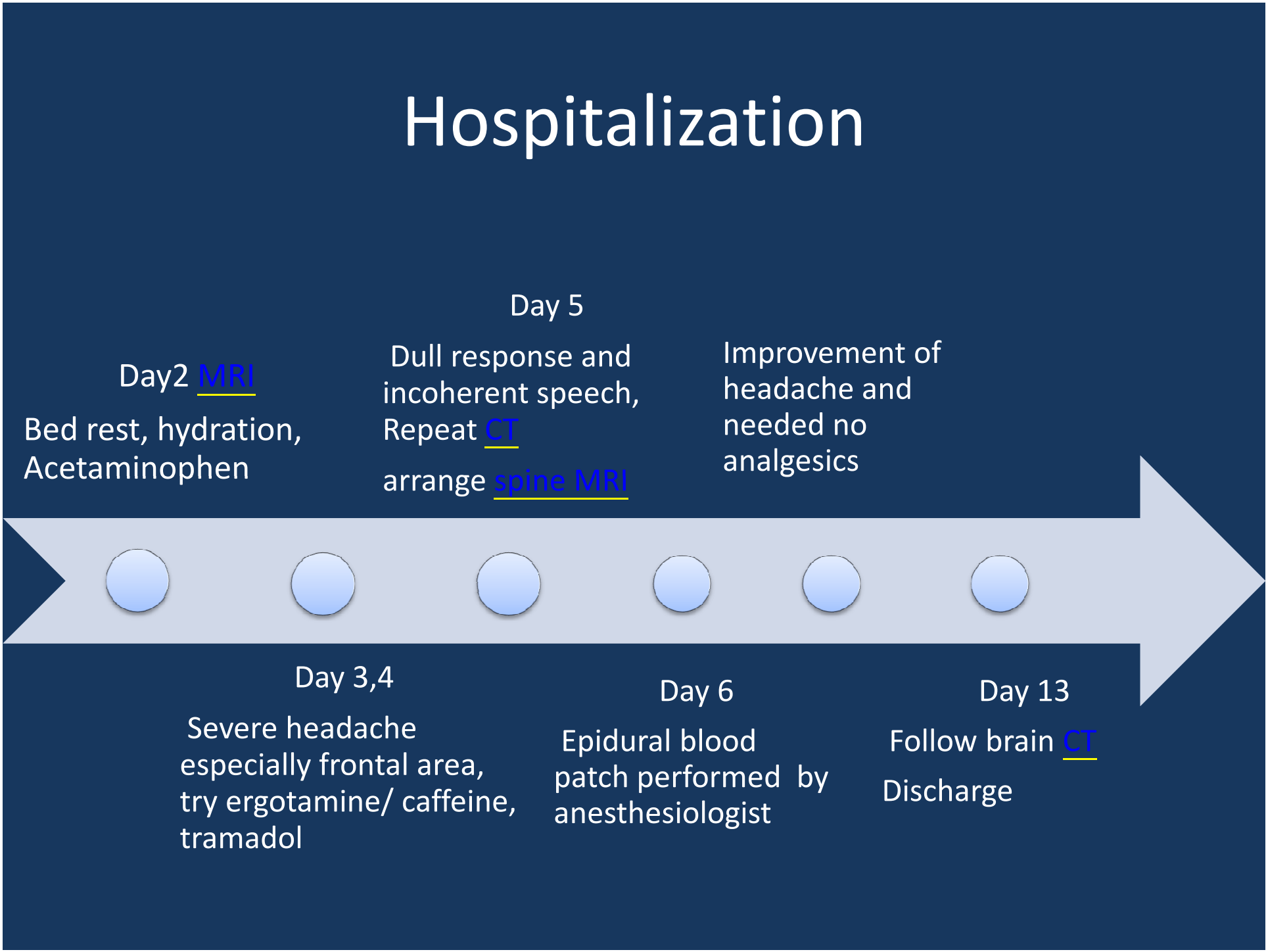
- Consciousness
  - Clear, cooperative

Neck supple

- **CN II**
  - Pupil size: 3.0 mm/ 3.0 mm (L/R)
  - Direct light reflex: +/+ (L/R) Indirect light reflex: +/+ (L/R)
  - Visual field: normal
  - Visual acuity: unable to be checked
  - Fundus: visible venous palsation, no papilledema
- **CN III, IV, VI**
  - EOM: no limitation

Others were all normal findings

Hospitalization



Spontaneous intracranial hypotension (SIH)

- 1938, Schaltenbrand, aliquorrhea.
- Annual incidence 5/100,000
- Female: Male=2:1
- Peak incidence: 40 years of age

- *The SIH should be considered an important cause of new daily persistent headaches, particularly among young and middle-aged patients.*

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Spontaneous intracranial hypotension (SIH)

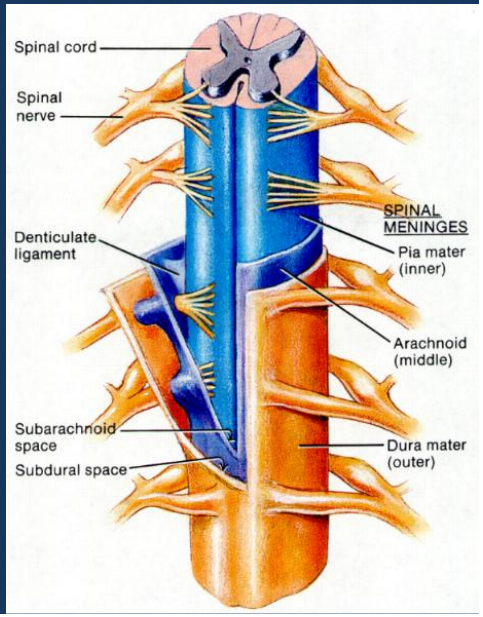
- Spontaneous spinal CSF leaks.
- Precise case remain unknown.

-> Traction on pain-sensitive intracranial and meningeal structure, particularly sensory nerves and bridging vein

-> underlying structural weakness of spinal meninges

-> Most CSF leaks at thoracic or cervicothoracic junction

- Traumatic event: chronic cough, lifting heavy objects.
- No risk of meningitis, unlike CSF rhinorrhea.
- Connective tissue disorder: Marfan syndrome, Ehlers-Danlos syndrome type II, autosomal dominant polycystic kidney disease.



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Diagnostic criteria (ICHD-II 7.2.3)-

Headache attributed to spontaneous(or idiopathic) low CSF pressures

A. Diffuse and/or dull headache that worsen within 15 minutes after sitting or standing, with at least one of the following and fulfilling criterion D:
   1. neck stiffness
   2. tinnitus(direct transmission CS
   3. hypacusia
   4. photophobia
   5. nausea

Variable pattern
Disappear orthostatic headache if leak no treated
Exertional headache
Occur at end of day
Paradixical headache, worsen when lying down
Intermittent headache

B. At least one of the following:
   1. evidence of low CSF pressure on MRI (eg, pachymeningeal enhancement)
   2. evidence of CSF leakage on conventional myelography, CT myelography, or cisternography
   3. CSF opening pressure < 60mmH2O in sitting position

C. No history of dural puncture or other cause of CSF fistula

D. Headache resolves within 72 hours after epidural blood patching

Commend

- The underlying disorder may be low CSF volume. A history of trivial increased in intracranial pressure (eg, on vigorous coughing) is often elicited. In other cases a *sudden drop in atmosphere pressure has occurred.*

- Many patients with spontaneous low CSF pressure headache *respond to epidural blood patching, epidural saline infusion* or pharmacological therapies such as *intravenous caffeine or* conventional analgesics. Some have spontaneous resolution of their headache, while others relapse after initial successful treatment.

- *Dural puncture should be avoided in patients with positive MRI signs.*

Table. Differential Diagnosis of Headache Due to Spontaneous Spinal Cerebrospinal Fluid Leak and Intracranial Hypotension*

Headache Disorder	Typical Age at Onset, y	Female-Male Ratio	Connective Tissue Disorders†	Headache Features	Thunderclap Headache	Associated Features	Confirmatory Testing
Primary							
New daily persistent headache	15-50 (Mean, 35)	2:1	No	Bilateral more common than unilateral Migraine or tension-type dominant	No	Nausea, fatigue, preceding viral illness	None
Secondary							
Spontaneous intracranial hypotension	20-60 (Mean, 40)	2:1	Yes	Bilateral much more common than unilateral Improved with recumbancy, but variable	Yes	Visual/aural changes, meningismus, cranial nerve dysfunction	MRI, LP, myelogram
Subarachnoid hemorrhage	>20 (Mean, 50)	1.5:1	Yes	Bilateral more common than unilateral Onset instantaneous in most, but may be gradual	Yes	Meningismus, cranial nerve dysfunction, seizure	CT, LP
Carotid/vertebral artery dissection	20-70 (Mean, 45)	1:1	Yes	Unilateral much more common than bilateral Often with neck pain	Yes	Horner syndrome, cranial nerve dysfunction, pulsatile tinnitus	Angiography
Cerebral venous sinus thrombosis	Any age	3:1	No	Bilateral more common than unilateral (but unilateral when headache is only sign)	Yes	Seizure, papilledema, visual changes, cranial nerve dysfunction	MRI/MRV, angiography
Benign intracranial hypertension	20-40 (Mean, 30)	8:1	No	Bilateral much more common than unilateral Worse in recumbancy	No	Papilledema, visual changes, abducens nerve palsy	LP
Posttraumatic headache	Any age	1.5:1	No	Bilateral much more common than unilateral Tension-type, often after mild head injury	No	Dizziness, neuropsychological symptoms	None
Meningitis	Any age	1:1	No	Bilateral	Yes	Fever, meningismus, systemic illness	LP

Not all orthostatic headache are caused by spontaneous CSF leaks, eg. postural bradycardia syndrome, Filum ependymoma

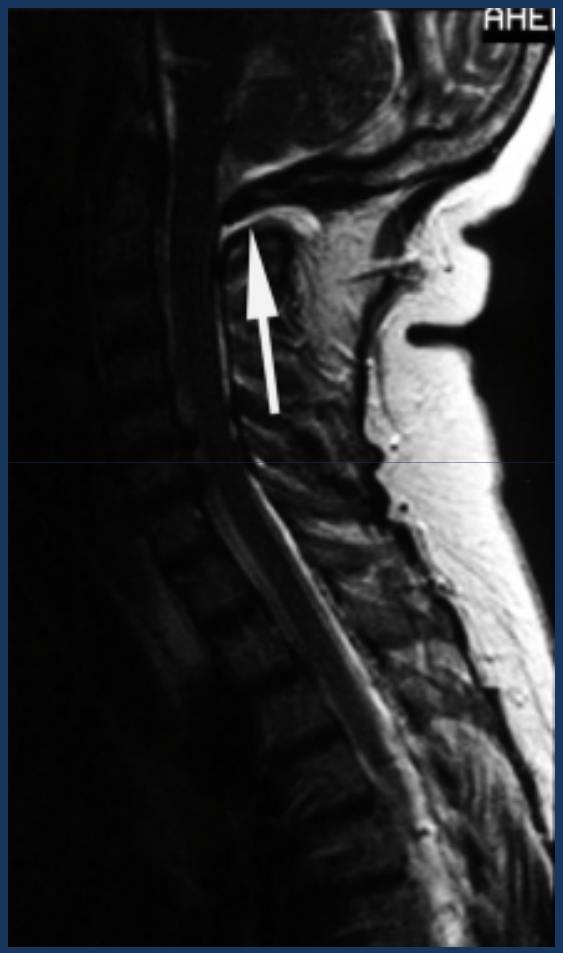
JAMA. 2006;295:2286-2296

MRI Findings

- Subdural fluid collections (50%)
- Enhancement of the pachymeninges (20%)
- Engorgement of cerebral venous sinuses
- Pituitary enlargement
- "Sagging" of the brain, cerebellar tonsillar herniation (>4.3mm) and descent of the brainstem
- Flattening of the optic chiasm, and increased anteroposterior diameter of the brainstem
- Decrease in the size of cisterns and ventricles

JAMA. 2006;295:2286-2296

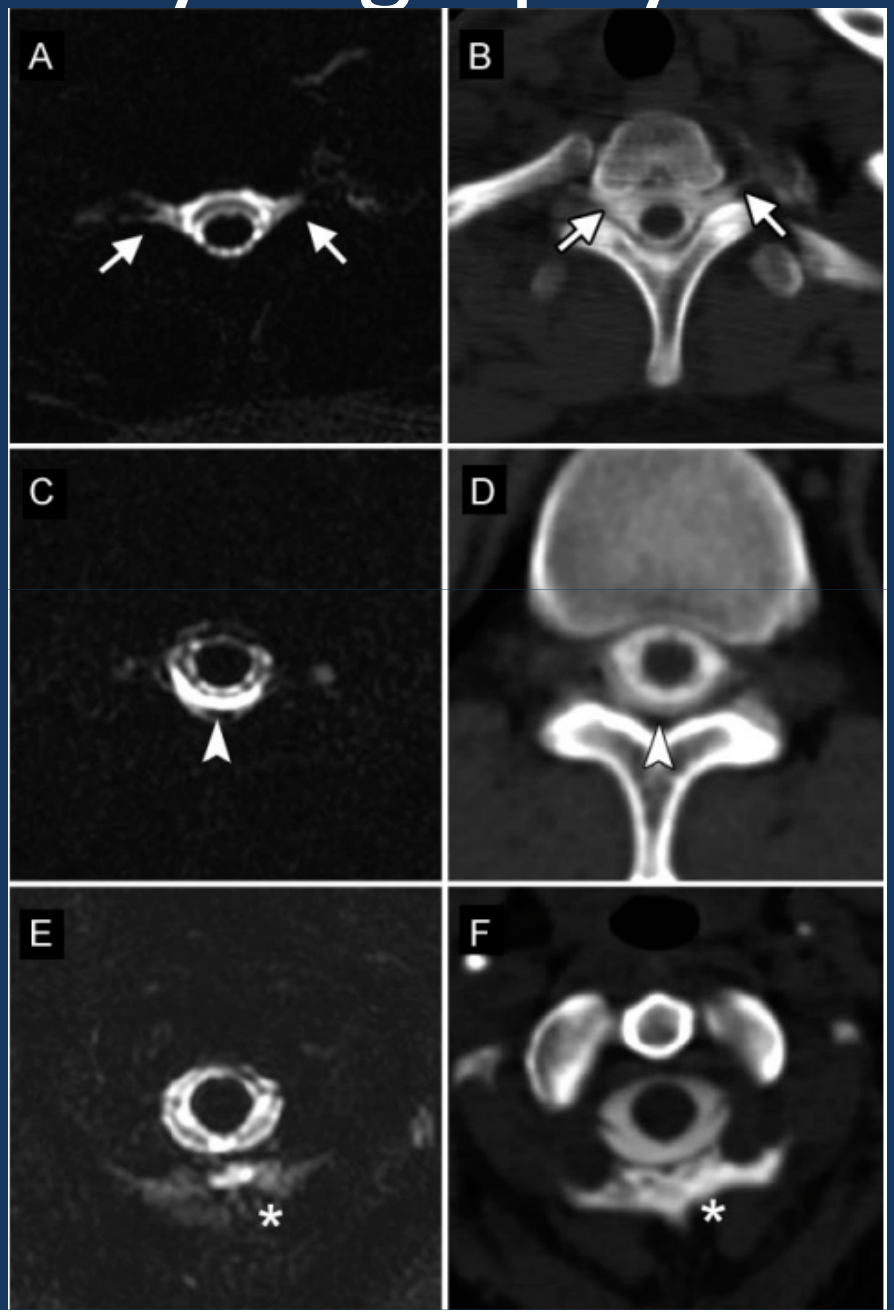
Spine MRI for CSF leak



Sagittal T2-weighted cervical MRI reveals a CSF leak at the C2 level

Sagittal T2-weighted cervical MRI showing CSF leak at the C1

Myelography



Magnetic resonance myelography (MRM; A) and computed tomographic myelography (CTM; B) showed bilateral CSF leaks at T1-2 (arrows) in a 24-year-old female patient. Axial images of MRM (C) and CTM (D) at T11-12 revealed posterior epidural CSF collections (arrowheads) in a 34-year-old male patient. High-cervical retrospinal CSF collections (asterisks) at C1-2 in a 28-year-old female patient were demonstrated on MRM (E) and CTM (F).

- ✓ MRM did not differ from CTM in the detection rates of CSF leaks along the nerve roots, high-cervical retrospinal CSF collections , and epidural CSF collections.
- ✓ MRM demonstrated more spinal levels of CSF leaks and epidural collections than CTM.
- ✓ Heavily T2-weighted magnetic resonance myelography was accurate in localizing CSF leaks for patients with spontaneous intracranial hypotension.
- ✓ This noninvasive technique may be an alternative to computed tomographic myelography before targeted epidural blood patches.

Neurology® 2009;73:1892–1898

Management of SIH

- Self-limited
- Conservative treatment

-> Hydration

-> Bed rest

-> Abdominal binder

- Intravenous steroid, caffeine, theophylline
- Epidural blood patch
- Surgical repair of CSF leak
- Intrathecal saline infusion until the CSF leak can be permanently repaired
- Osmotic or loop diuretics are not effective

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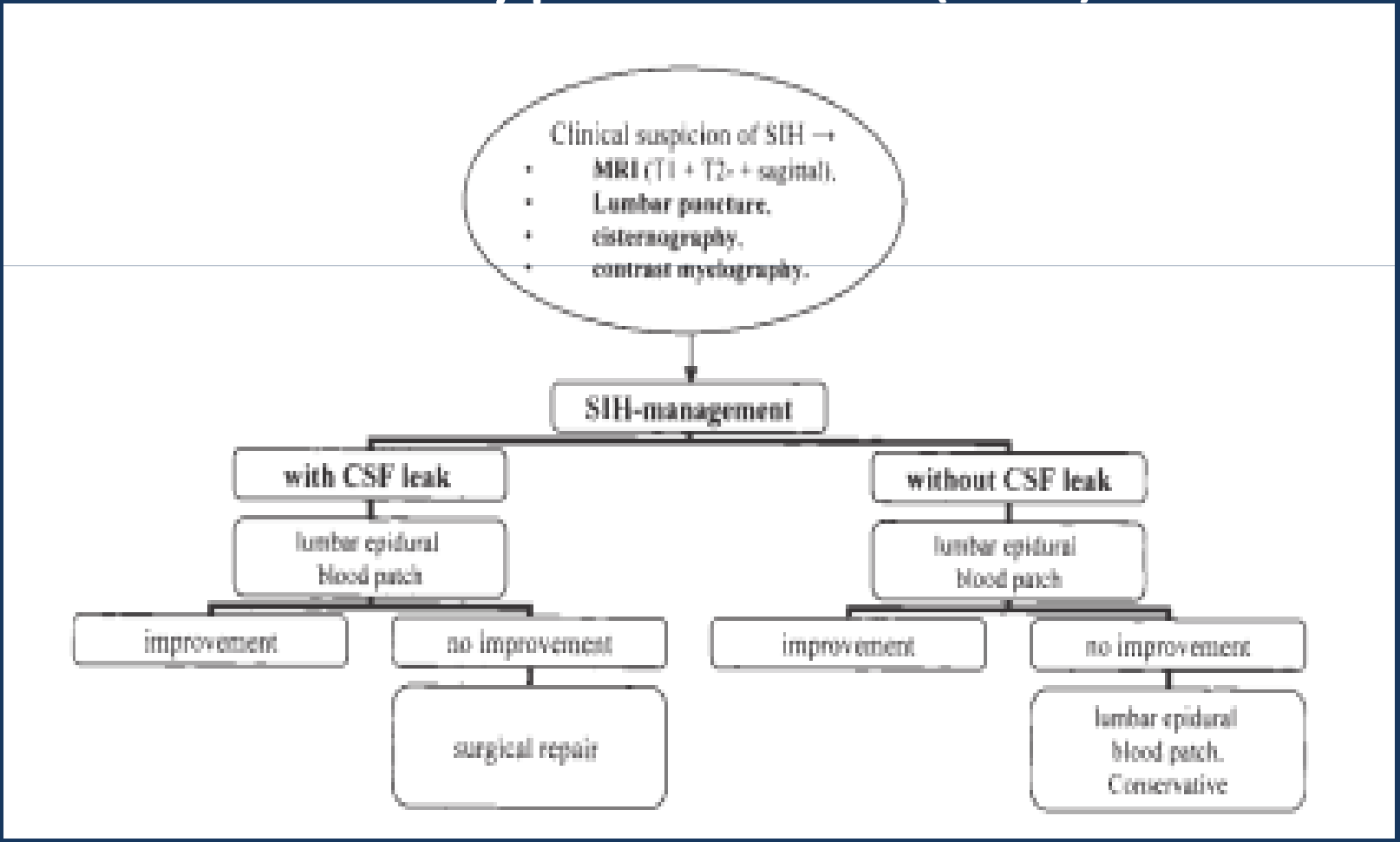
Algorithm of diagnosis and management of spontaneous intracranial hypotension (SIH)



J. Neurosurg. / Volume 96 / May, 2002

Complication of SIH

- Subdural fluid collection
- Subdural hematoma
- Cranial nerve palsy (eg. abducens palsy)
- Cerebral sinus venous thrombosis

@ The diagnosis of SIH should be considered in young patients presenting with SDH in the absence of trauma and with normal clotting.

Cephalalgia 2007; 27:133–138

Subdural hematoma or subdural fluid collection in SIH

- Rupture of bridging veins pulled away from the dura due to low intracranial pressure and brain descent.
- Fragile and dilated new dural vessels => delayed SDH
- There is no consensus regarding the management of SDH cause by SIH.
- Surgical indication (as chronic SDH)

-> Focal neurologic deficits
-> Decreased level of consciousness
-> SDH with maximal thickness >1cm

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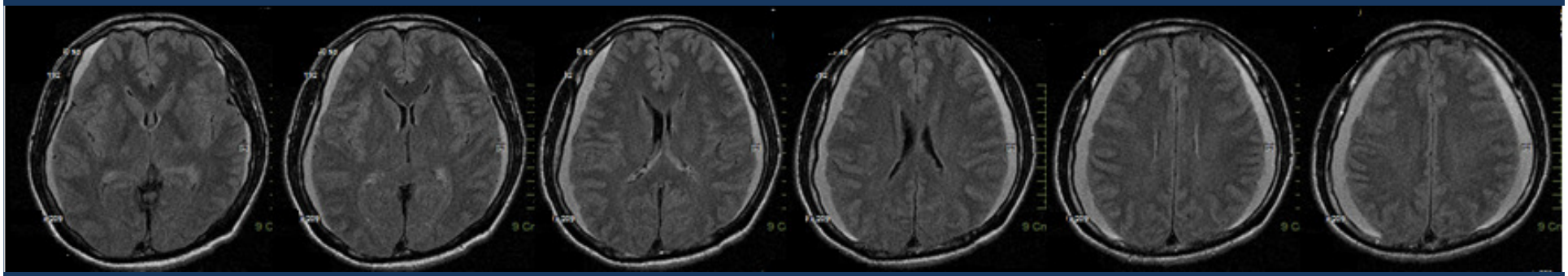
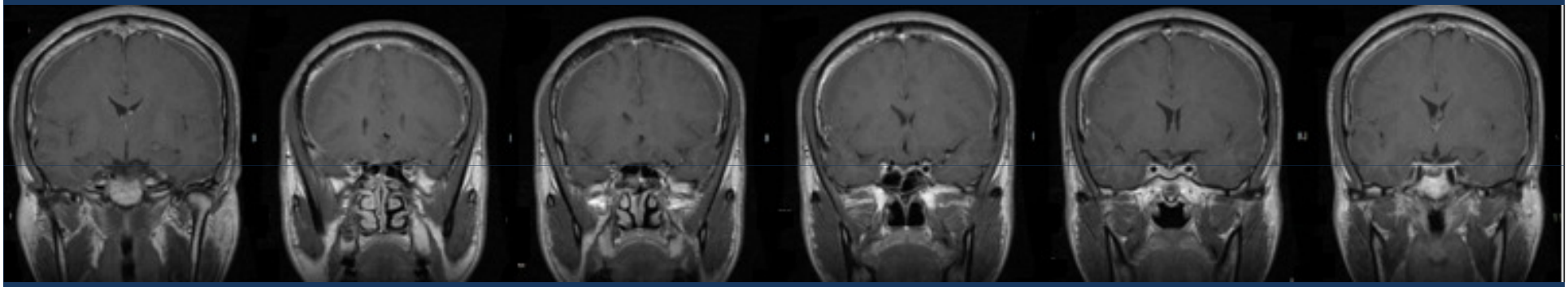
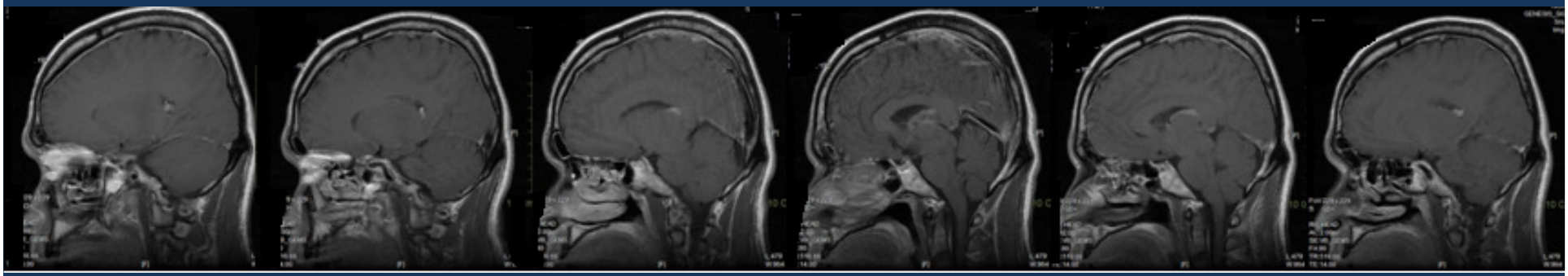
Cephalalgia 2007; 27:133–138

Discussion







11/11 brain CT