Intracranial hypotension syndrome

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Learning objectives

- Review of the pathophysiology and clinical data intracranial hypotension syndrome (SHI).
- Show the imaging findings useful in the diagnosis of SHI through case studies of our institution.
- Ask differential diagnosis of diseases presenting with similar radiological findings.
Background

Intracranial hypotension (IH) is a syndrome that is characterized by postural headache associated with low cerebrospinal fluid (CSF) pressure (either spontaneous or secondary) or CSF leakage.

It's a benign condition of difficult diagnosis because it's highly variable and nonspecific clinical and imaging presentation. The broad spectrum of symptoms including nausea, vomiting, neck pain, vertigo, visual and hearing disturbances [2-3].

The diagnostic criteria and classification according The International Classification of Headache Disorders (ICHD), 3er edition, are:

**Diagnostic criteria:**

A. Any headache fulfilling criterion C

B. Low CSF pressure (<60 mm CSF) and/or evidence of CSF leakage on imaging

C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or led to its discovery

D. Not better accounted for by another ICHD-3 diagnosis.

**Classification:**

- Post-dural puncture headache (Headache within 5 days after puncture that remits spontaneously within 2 weeks, or after sealing of the leak with autologous epidural lumbar patch)

- CSF fistula headache

- Headache attributed to spontaneous intracranial hypotension (SIH). SIH cannot be diagnosed in a patient who has had a dural puncture within the prior month. It is presented around de 5 per 100,000 persons with female preponderance in a ratio de 2:1 [2-3,6]. Generalized connective tissue disorders (Marfan’s Syndrome, Ehler Danlos Syndrome type II, autosomal dominant polycystic disease) ARE suspected up to two thirds of patients [2-3,6].
IH is the result of the passage of CSF of subarachnoid space to epidural space by a defect in dura, which is not always known. The exit to the epidural space of CSF makes decrease pressure and volume intracranial, causing reduction of the buoyant force and downward displacement of the brain with traction of cranial nerves and pain sensitive structures. Traction of these structures explains some of clinical symptoms. Also, the intracranial content (blood, brain, CSF, meninges and extracellular fluid) in intact skull must remain constant as described in the Monro-Kellie hypothesis and a loss of CSF volume must be compensated by increased extracellular fluid and/or blood volume [2-3,5-6,8].

The MR findings associated with IH can be predicted with Monro-Kellie hypothesis and the downward displacement of the brain.
Findings and procedure details

MR is an important tool in the diagnosis of intracranial hypotension, several imaging findings have been described, however they are variable and may not always be present.

MR imaging include:

**QUALITATIVE:**

**Cranial MR imaging:**

- **Pachymeningeal enhancement after contrast administration that may be localized or diffused resulting in vasocongestion and interstitial edema in the dura mater by prolonged intracranial hypotension.** The Pachymeningeal enhancement is thick linear, no Leptomeningeal enhancement (sulci or brain surface), enhancement above and below the tentorium (Figs. 1, 2) [1-8].

  There may be misdiagnosis with neoplastic, inflammatory, or granulomatous disease.

- **Subdural collections:** The mechanism is unknown. One explanation is the direct leakage of CSF into the subdural space. Another theory is the rupture of small capillaries because of the downward displacement of the brain (Fig.3).

- **Enlargement of the pituitary gland** occurs in IH secondary to dilation of the venous side of the circulation (Fig. 4).

- **The venous distension sign (VDS).** Dural sinuses enlarge as they compensate for the loss intracranial CSF volume. The expansion results in a characteristic change in the contour of the dural sinuses. This contour change significantly at the level of the inferior border (normally it's concave or straight) of the dominant transverse sinus visualized in its mid-portion on sagittal images of the brain. The overall sensitivity of the VDS for the diagnosis of intracranial hypotension was 94% and specificity was 94% [3-6] (Fig. 5).

- **Brain sagging** is very specific finding in IH. It has several features, such as effacement of the suprasellar cistern, bowing of the optic chiasm over the pituitary fossa, flattening of the pons against the clivus, and descent of the brain (low cerebellar tonsils, the iter in the aqueduct and the splenium corporis callosi are displaced downward more of 2 mm below the incisural line connecting the tuberulum sellae and the entrance of the vein of Galen into the straight sinus) (Fig. 6).
Spinal MR imaging:

- **Fluid collections.** The collections tended to be non-focal and often extended over five or more spinal segments. Axial T2 weighted images were useful to assess the dura deep to the collection, finding that confirms epidural location (Fig. 7).

- **Dilated epidural veins.** The presence of dilated veins may be explained by the Monro-Kellie hypothesis.

- **Thickening and festooned morphology of epidural venous plexus.**

- **Dural enhancement is** result from dural vasodilatation and engorgement.

**QUANTITATIVE [2]:**

- **Pontomesencephlic angle** # 55°. It is defined as the angle between a line tangential to the anterior margin of the midbrain and the line tangential to the superior margin of the pons (Fig. 8).

- **Mamillopontine distance** # 5 mm. It is defined as the distance between the inferior aspect of the mammillary bodies and the superior aspect of the pons. This measurement approximates the interpeduncular cistern (Fig. 9).
Fig. 1 Diagram illustrates pachymeningeal enhancement (orange line). Which is present adjacent to inner table of the skull, in the falx within interhemispheric fissure, and in the tentorium.

Fig. 1

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Fig. 2 Axial gadolinium-enhanced T1 weighted magnetic resonance imaging demonstrates diffuse pachymeningeal enhancement (arrows).

Fig. 2

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Fig. 3 Axial T2 weighted image show subdural collection in 31 year old woman with spontaneous intracranial hypotension (arrow).

Fig. 3

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Fig. 4 Axial T1 weighted post contrast: enlargement of the pituitary gland (arrows).

Fig. 4

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Fig. 5 Sagittal T1 weighted images with (A) and without (B) contrast-enhanced through middle third of the dominant transverse sinus show convex bulging of its inferior border (arrows).

**Fig. 5**

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Fig. 6 Sagittal T1 weighted image show flattening of the pons (arrows).
Fig. 7 Woman with spontaneous intracranial hypotension (A) and control image 5 month later (B). A, Sagittal T1 weighted image show ventral displacement of cord and dorsal extradural fluid collection (arrows). B, Sagittal T1 weighted image show practical resolution of dorsal extradural fluid collection (arrows).

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Fig. 8 Sagittal T1 weighted image. Pontomesencephalic angle is the angle between line drawn along anterior margin of midbrain (green line) and anterior superior margin of pons (orange line).

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Fig. 9 Sagittal T1 weighted image. Mamilllopontine distance is the distance between inferior aspect of mamillary bodies to superior aspect of pons (red line).

Fig. 9

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Conclusion

Headache is one of the most frequent causes of medical consultation, IH is a cause benign and rare, commonly misdiagnosed, causing delay in effective treatment and exposing patients to risks from treatments for other pathologies that mimic IH. To know and understand the qualitative and quantitative imaging findings help to make a more accurate diagnosis.
References


