17 Intracranial Hypotension

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INTRODUCTION

Intracranial hypotension is a clinical manifestation of low cerebrospinal fluid (CSF) volume or pressure caused by a dural CSF leak. It is usually precipitated by dural injury from an iatrogenic event such as a lumbar puncture, recent surgery, overshunting, or trauma.

Spontaneous intracranial hypotension (SIH) was first described by Georg Schaltenbrand in 1938[1] and has been recognized as a rare but significantly underdiagnosed differential consideration for severe headache.[2] Although its true incidence is unknown, a few observational studies have estimated its prevalence as 2 to 5 in 100,000 patients with primary headache.[2,3] In these studies, female cases were twice as frequent as male cases, with a peak incidence in middle age.[2,3]

The classic clinical presentation symptom of intracranial hypotension is orthostatic headache shortly after lumbar puncture, craniospinal surgery, or trauma. Orthostatic headache is also the most frequent presenting symptoms of SIH.[2] The pattern of headache is usually diffuse, with varying degrees of severity and acuity. Other less sensitive and nonspecific symptoms include neck pain/stiffness, nausea, tinnitus, hyperacusis, and photophobia. In rare cases, severe brain displacement from intracranial hypotension can precipitate coma or stupor.[4] Subdural hematoma and venous sinus thrombosis are sequelae of intracranial hypotension but can be the presenting symptoms in unsuspected SIH cases.[5,6] Diagnostic criteria for SIH have been established by the International Headache Society (Box 17.1).[7,8]

EVOLUTION: OVERVIEW

Intracranial hypotension is a direct consequence of dural CSF leakage. The cause and location of CSF leakage may be obvious in iatrogenic or traumatic cases even without imaging investigations (e.g., CSF rhinorrhea or otorrhea). However, the specific location of the dural defect leading to SIH is often unknown. Computed tomography (CT) myelography, and intrathecal gadolinium magnetic resonance (MR) myelography are the most sensitive imaging modalities for localizing spinal CSF leaks and are preferred over radionuclide cisternography, given the latter's low spatial resolution. Although there is a theoretical concern of cerebral herniation in performing a myelogram, there have been no reported cases of this and the expected low cerebral CSF pressure is also reassuring.[2]

As can be inferred from the diagnostic criteria in Box 17.1, magnetic resonance imaging (MRI) of the brain plays a major, noninvasive role in the evaluation of intracranial hypotension. Brain MRI can confirm suspected iatrogenic/traumatic cases by demonstrating positive imaging features while ruling out other etiologies to account for the patient's symptoms. The characteristic MRI findings of intracranial hypotension (Fig. 17.1) are listed in Box 17.2 and include diffuse smooth pachymeningeal enhancement (80%–100% sensitivity), subdural fluid collections/hematomas (27%–69% sensitivity), venous engorgement (75% sensitivity), pituitary hyperemia (unknown sensitivity), and brain sagging (10%–62% sensitivity).[2,3,9-11] Brain sagging or downward displacement is a highly specific MRI feature, including flattening of the pons against the clivus, effacement of the prepontine or perichiasmatic cisterns, and descent of the cerebellar tonsils.

In some severe or chronic cases of intracranial hypotension, pachymeningeal thickening/enhancement can be prominent and may even involve the spinal dura (Fig. 17.2).

Many authors argue that intracranial hypotension is not an issue of decreased intracranial pressure as the name suggests but of the loss of intracranial CSF volume. This was initially suggested based on observations that some patients with classic clinical symptoms and positive imaging finding were found to have normal CSF opening pressures.[2,3,10,11] The claim was further supported by multiple studies failing to demonstrate a correlative relationship between CSF opening pressure and imaging findings.[2,11,12]

Low CSF volume can explain the pathophysiology of MRI findings on the basis of the Monroe-Kellie doctrine (Fig. 17.3A). Loss of CSF volume is compensated by increasing intracranial vascular volume to a greater extent in the venous compartment than in the arterial compartment owing to its lower pressure as well as the large intracranial venous capacity.[2,13] This augmented blood volume will manifest on contrast-enhanced MRI as venous engorgement, smooth pachymeningeal enhancement, and pituitary

BOX 17.1 Criteria for Diagnosing Spontaneous Intracranial Hypotension

A. Orthostatic headache
B. The presence of at least one of the following:
   - Low CSF opening pressure (<6 cm H_2O)
   - Sustained improvement of symptoms after epidural blood patching
   - Demonstration of an active spinal CSF leak
   - Cranial MRI findings of intracranial hypotension
C. No recent history of dural puncture
D. Not attributable to another disorder

CSF, Cerebrospinal fluid; *MRI*, magnetic resonance imaging.
Modified from Schievink WI, Dodick DW, Mokri B, et al. Diagnostic criteria for headache due to spontaneous intracranial hypotension: a perspective. ***Headache****. 2011;51:1442–1444.*

BOX 17.2 Brain Magnetic Resonance Imaging Findings of Intracranial Hypotension

- Diffuse, smooth pachymeningeal enhancement
- Subdural hygroma/collection/hematoma
- Intracranial venous engorgement
- Pituitary hyperemia
- Brain sagging or downward displacement
  - Flattening of the pons against the clivus
  - Effacement of prepontine or perichiasmatic cisterns
  - Cerebellar tonsil descent

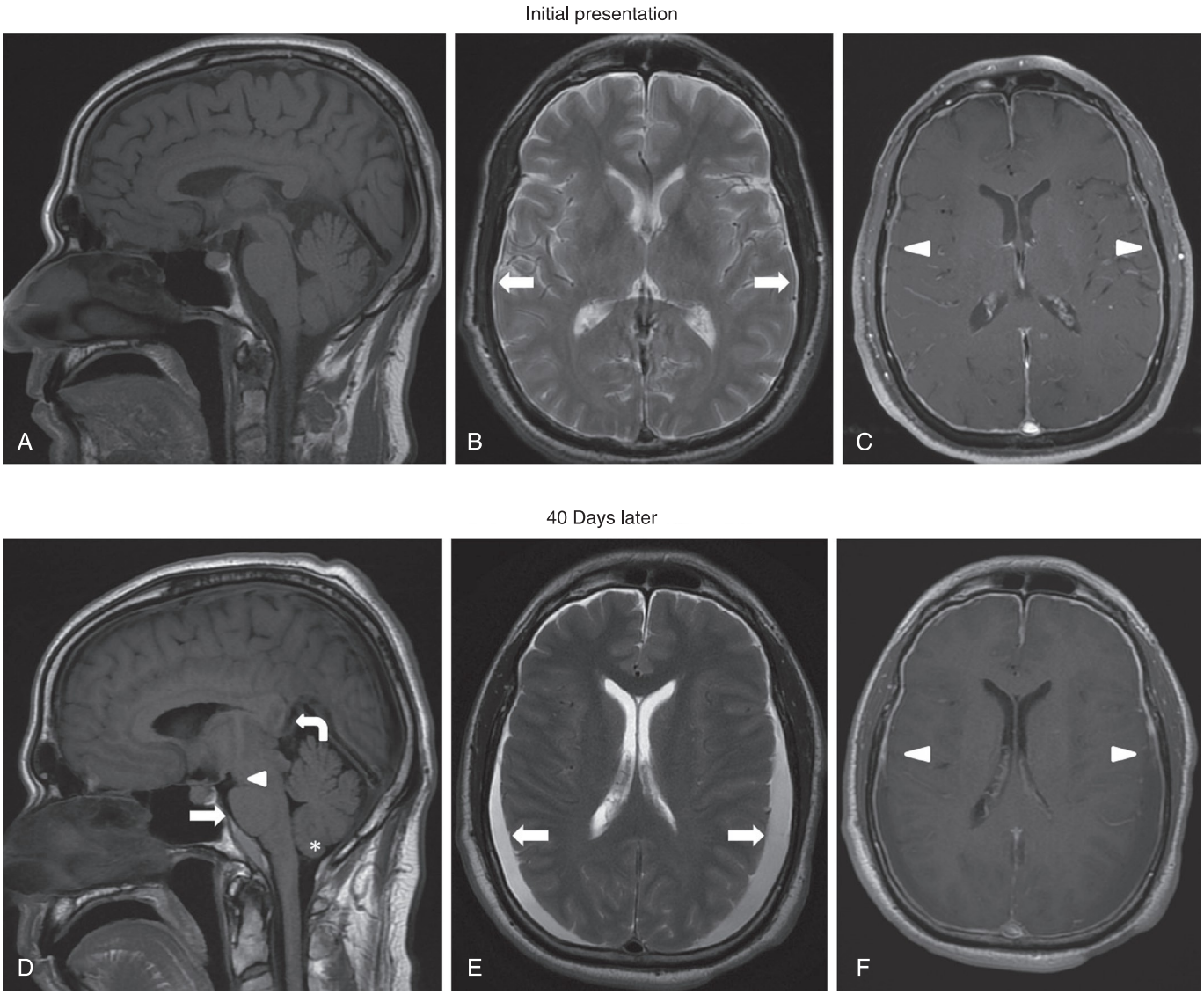

Figure 17.1. Natural history of intracranial hypotension with subsequent subdural hematoma formation. Evolution of magnetic resonance imaging (MRI) findings in a patient with intracranial hypotension due to cervical dural cerebrospinal fluid leak precipitated by osteophytes. The upper MRI sequence was acquired at the time of initial presentation; due to worsening clinical symptoms, the lower sequence was obtained 40 days after initial imaging. Sagittal T1-weighted images of the brain (D) reveal interval downward brain sagging compared with initial imaging (A): flattening of pons against clivus and effacement of the prepontine cistern *(arrow)*, downward sagging of corpus callosum *(curved arrow)*, narrowed midbrain-pons angle *(arrowhead)*, and subtle cerebellar vermis descent *(asterisk)*. T2-weighted axial images (B and E) demonstrate initial extra-axial spaces commensurate with dural thickening (B, *arrows*), with interval development of prominent bilateral subdural collections (E, *arrows*). T1-weighted postcontrast axial images (C and F) show diffuse, smooth pachymeningeal enhancement with mildly increased in thickness overlying the frontal lobes *(arrowheads)*.

hyperemia (see Fig. 17.3B). Of note, this enhancement pattern represents prominence of the extraparenchymal vascular volume outside of the blood-brain barrier (BBB) rather than pathologic disruption of the BBB. Venous engorgement and loss of CSF pressure may impede normal CSF resorption, which manifests typically as thin, diffuse subdural hygromas without significant mass effect.[2,5] Subsequently, increased subdural space pressure stretches engorged bridging subdural veins, eventually resulting in tearing of these veins and subdural hemorrhage. Last, brain sagging is hypothesized to happen due to direct loss of buoyancy as a consequence of decreased intracranial CSF volume.

It is well recognized that despite extensive imaging, no epidural CSF leak is found in many cases of SIH. In only a minority of patients with SIH will a myelogram localize the CSF leak site in the form of a small dural defect or even a CSF-venous fistula,[14] whereas a slow and intermittent CSF leak may not be evident (see Fig. 17.3C).[15] Furthermore, the leak site can be difficult to establish in large leaks, as extensive myelographic contrast extravasation may occur prior to imaging.

The association of nerve root sleeve diverticula with SIH remains a controversial topic. Although diverticula are frequently found near the site of contrast leakage, the vast majority are incidental findings of no clinical significance in asymptomatic patients. There is an association between SIH and generalized connective tissue disorders such as Marfan and Ehlers-Danlos disease and autosomal dominant polycystic kidney syndrome (ADPKD), although the exact pathophysiologic relationship is not proven. Finally, there are

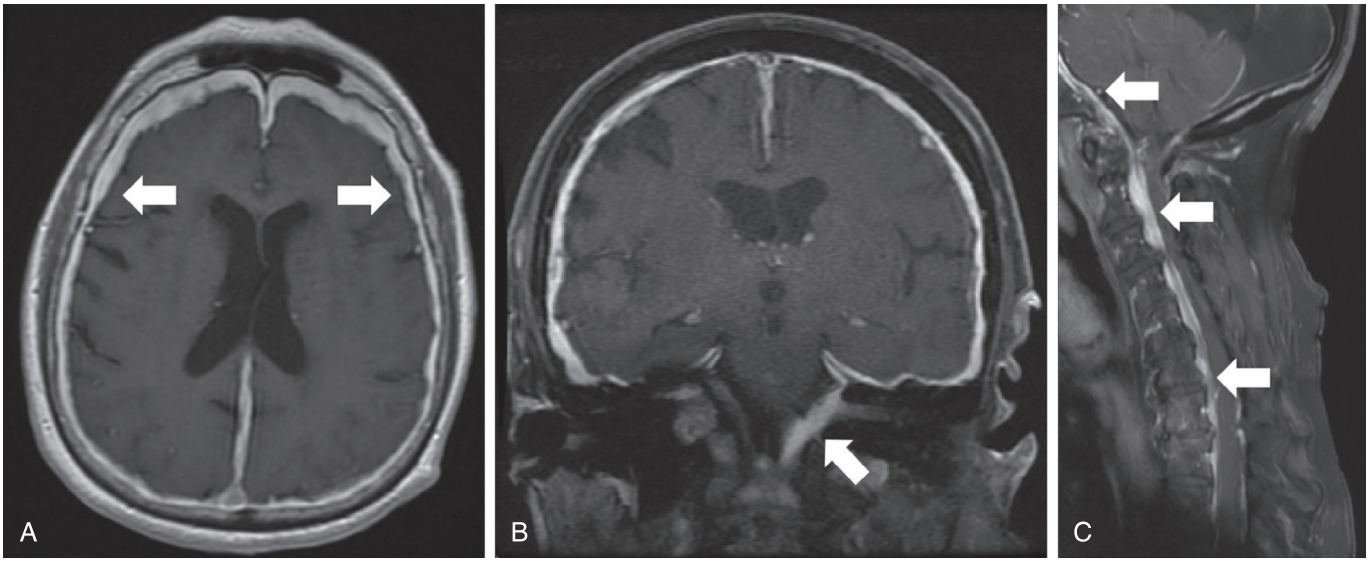

Figure 17.2. Marked chronic pachymeningeal thickening and enhancement due to chronic overshunting. T1 postcontrast images in a patient with long-standing ventricular-peritoneal shunt and chronic overshunting resulted in marked smooth pachymeningeal thickening and enhancement throughout the neural axis, including supratentorial (A, *arrows*), infratentorial (B, *arrow*), and spinal canal (C, *arrows*) involvement.

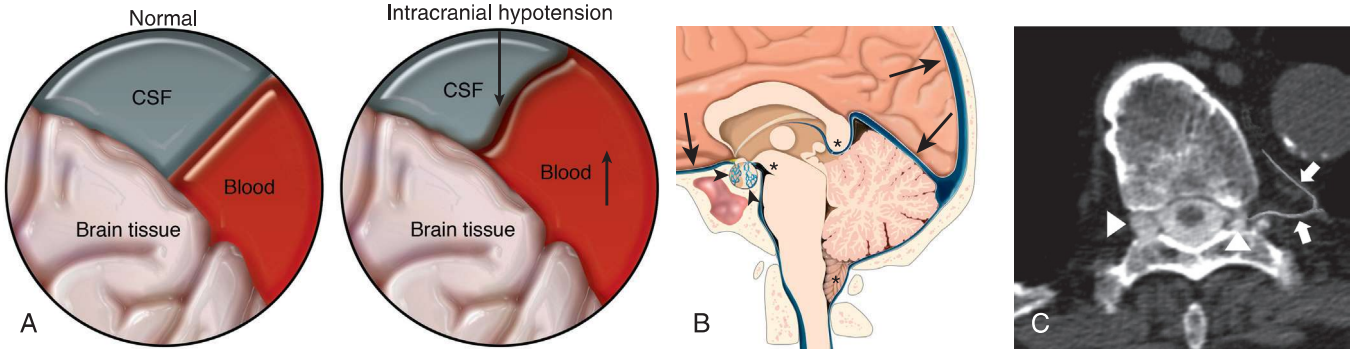

Figure 17.3. (A) According to the Monroe-Kellie doctrine, loss of cerebrospinal fluid *(CSF)* volume is compensated by increasing intracranial vascular volume to a greater extent in the venous compartment than the arterial compartment due to its lower pressure as well as the large intracranial venous capacity. (B) Augmented blood volume will manifest on contrast-enhanced magnetic resonance imaging scans as venous engorgement *(arrows)*, smooth pachymeningeal enhancement, and pituitary hyperemia *(arrowheads)*. Brain sagging *(asterisks)* is due to loss of buoyancy from decreased intracranial CSF volume. (C) Illustration of CSF venous fistula. In a CSF–venous fistula, a direct communication between the spinal subarachnoid space and an adjacent paraspinal vein permits loss of CSF into the venous circulation. Computed tomography myelogram images may reveal a "hyperdense paravertebral vein" sign[4] *(arrows)* indicative of intrathecal contrast material draining into a paravertebral vein. A high percentage of CSF–venous fistulas (>80%) are associated with a nerve root sleeve diverticulum *(arrowheads)*, implying a pathophysiologic connection.

reported cases of SIH precipitated by spinal degenerative changes including osteophytes or herniated disc material.[16,17]

EVOLUTION: IN GREATER DEPTH

Except for brain sagging, none of the MRI features in Box 17.2 is highly specific as an isolated finding. In addition, approximately 20% of intracranial hypotension cases demonstrate normal MRI findings.[3,8] When approaching chronic headache cases, the imaging interpreter should keep in mind that varying combinations of multiple subtle imaging features are more specific than a single dominant finding. Some authors attribute largely underdiagnosed cases of SIH to the subtle, variable nature of MRI findings.[2,11]

Upon successful treatment of a CSF leak, reversibility of brain MRI findings has been observed (Fig. 17.4). Improvement of imaging findings usually lags behind the clinical improvement in a range of a few days to weeks.[2,12,18]

MIMICS AND DIFFERENTIAL DIAGNOSIS

As mentioned previously, any isolated finding of MRI features listed in Box 17.2 can be misleading, and an interpreting physician should carefully look for relevant history and investigate other signs of intracranial hypotension before concluding the case.

Intracranial hypotension can be easily misdiagnosed as Chiari 1 malformation (Fig. 17.5), as these entities share common imaging findings of cerebellar tonsil descent in the context of a similar clinical presentation.[19] Furthermore, pituitary enhancement can mimic pituitary tumor[9] and subdural hemorrhage can be falsely attributed to trauma or a postsurgical complication.

Lastly, pachymeningeal enhancement has a broad differential diagnosis including metastatic disease, inflammatory and infectious meningitis, and autoimmune disease (Fig. 17.6). Care must be taken to confirm a smooth, diffuse pattern of pachymeningeal enhancement, especially when this finding is not accompanied by other MRI features of intracranial hypotension.

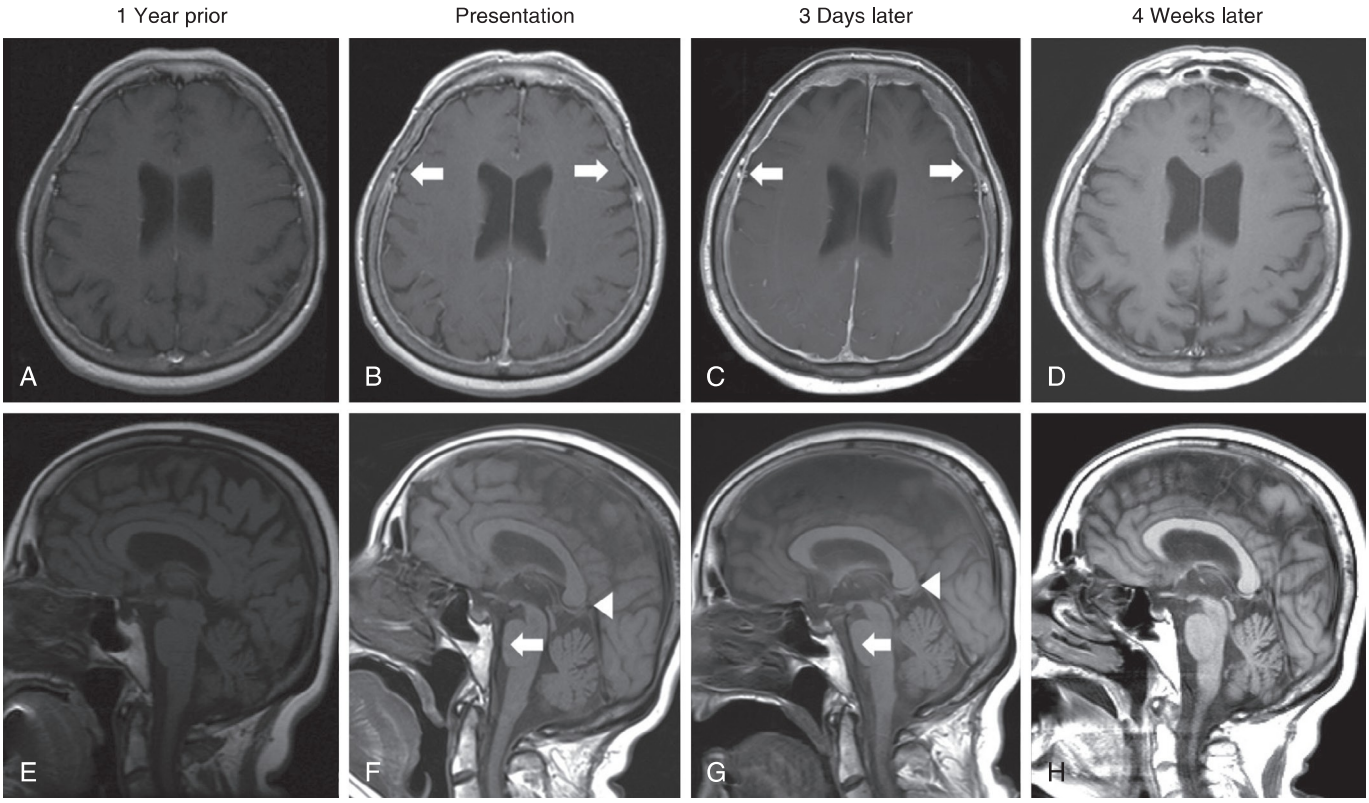

Figure 17.4. Longitudinal evolution of magnetic resonance imaging (MRI) findings in spontaneous intracranial hypotension. Approximately 1 year prior to presentation, the patient had a normal brain MRI (A and E). At presentation, the patient had an acutely worsening headache, at which time MRI images (B and F) demonstrate thin, smooth pachymeningeal enhancement (B, *arrow*) as well as midbrain flattening (F, *arrow*) and sagging of the corpus callosum *(arrowheads)*. Three days after the initial presentation (C and G), a short-interval follow-up MRI reveals thicker pachymeningeal enhancement (C, *arrow*) and persistent brain sagging (G, *arrow*). Although no clear etiology was identified after an extensive workup, the patient's symptoms and MRI findings spontaneously and completely resolved 4 weeks later (D and H).

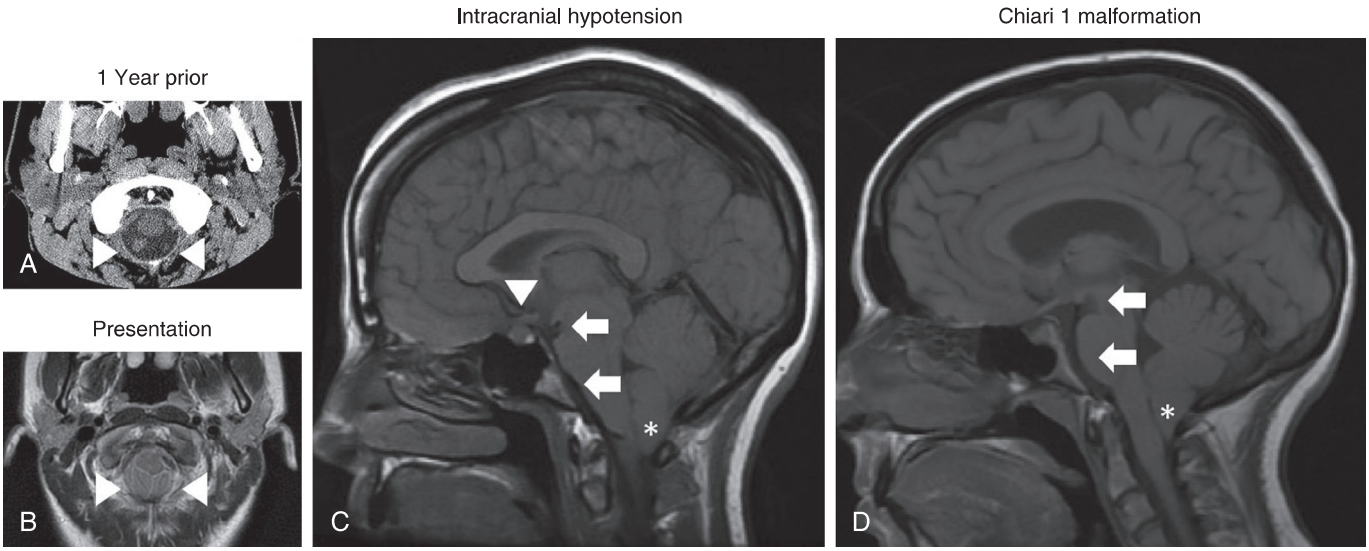

Figure 17.5. Intracranial hypotension (A to C) versus Chiari 1 malformation (D). Axial computed tomography (A) and T2-weighted magnetic resonance imaging (MRI) sequence (B) at the level of the foramen magnum with a 1-year interval reveals interval development of cerebellar tonsillar descent and crowding of the foramen magnum *(arrowheads)*. A T1-weighted sagittal image of the same patient at presentation (C) demonstrates typical MRI findings of intracranial hypotension: pituitary engorgement *(arrowhead)*, midbrain/pons downward sagging *(arrows)*, as well as cerebellar tonsil descent *(asterisk)*. A T1-weighted sagittal image of a different patient with Chiari 1 malformation (D) demonstrates the classic "peg-like" appearance of the inferior cerebellar tonsils *(asterisk)* but a normal-appearing brainstem *(arrows)*.

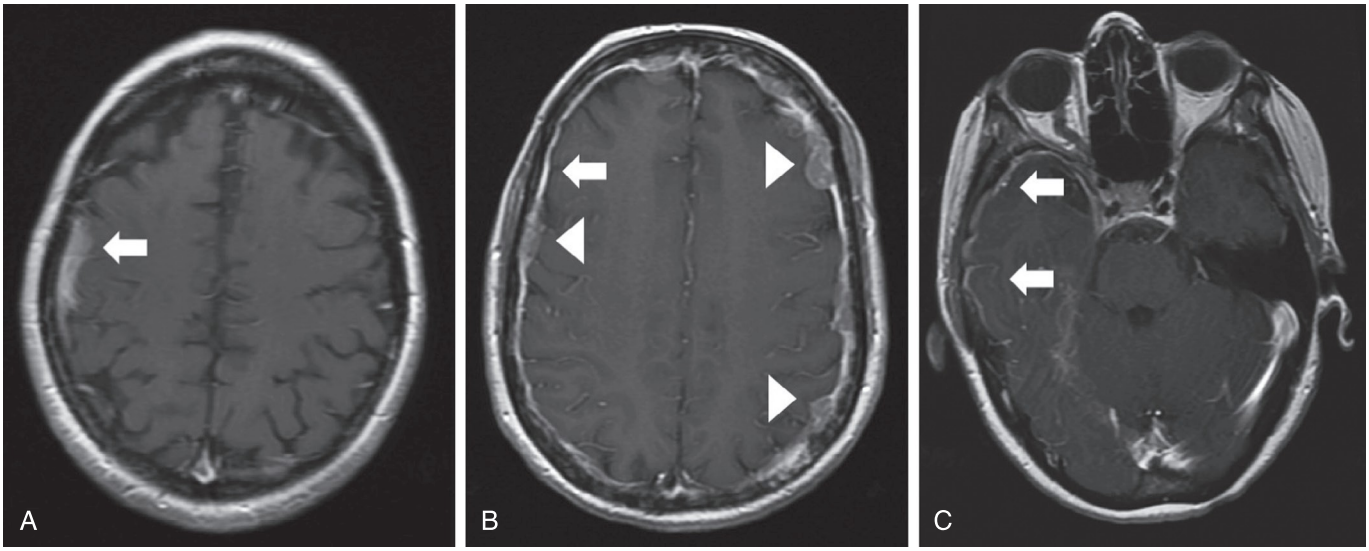

Figure 17.6. Mimics of pachymeningeal enhancement. (A) Postcontrast T1-weighted axial image of a patient with non-Hodgkin lymphoma reveals a focal area of pachymeningeal enhancement *(arrow)*. (B) Postcontrast T1-weighted axial image of a patient with diffusely metastatic breast cancer demonstrates pachymeningeal enhancement *(arrow)* with intervening focal nodular masses *(arrowheads)*. (C) Postcontrast T1-weighted axial image of a patient with focal bacterial meningitis from right otomastoiditis shows a focal area of asymmetric, smooth right temporal dural and leptomeningeal enhancement *(arrows)*.

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