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Case report

Intracranial hypotension secondary to spinal pathology: Diagnosis and treatment

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ABSTRACT

Spinal pathology resulting in cerebrospinal fluid (CSF) leak and intracranial hypotension is an infrequently reported and a potentially severe cause of headaches. We present a case of cerebrospinal fluid (CSF) leak caused by a thoracic disk herniation successfully treated with two targeted epidural blood patches. Although patients typically present with orthostatic headaches, the imaging findings of intracranial hypotension should prompt investigation of the spine for site and cause of the CSF leakage. Treatment includes autologous blood patch and surgery in refractory cases.

1. Introduction

Intracranial hypotension from cerebrospinal fluid (CSF) leak is a rare and potentially severe cause of refractory headaches. The condition can be a diagnostic challenge since the chief compliant is headaches while its etiology is elsewhere in the neural axis. Often the CSF leak occurs spontaneously from a suspected inherent weakness in the dura and is referred to as the syndrome of spontaneous intracranial hypotension (SIH) [1,2]. However, spinal pathology such as bone spurs and disc herniations can cause mechanical tears in the dura resulting in significant CSF leakage and intracranial hypotension [1]. While there have been a few case reports of CSF leaks secondary to spinal pathology, majority of the published literature on intracranial hypotension has focused on the diagnosis and treatment of spontaneous CSF leaks. In this paper, we present a review of the world literature on CSF leaks secondary to spinal pathology with a focus on treatment and outcomes.

2. Methods

A pubmed search using the key terms intracranial hypotension, syndrome of intracranial hypotension, CSF leakage, and spine was performed. Literature discussing intracranial hypotension secondary to CSF leakage from spinal pathology from 1990 to 2015 was selected for review.

2.1. Case report

A 41 year old female with past medical history significant for obesity, gastric bypass surgery, chronic anemia, and cholecystectomy presented to the emergency department with a ten day history of worsening positional headaches, photophobia, nausea, and vomiting. The symptoms were spontaneous without triggering event or history of trauma. Magnetic resonance imaging (MRI) of the brain demonstrated classic signs of intracranial hypotension including smooth pachymeningeal enhancement and downward displacement of the cerebellum (Fig. 1). Computed tomography (CT) myelography of the entire neural axis was then performed which demonstrated extradural extravasation of contrast consistent with CSF leak associated with a T5-T6 disc herniation (Fig. 2). The disc herniation appeared sharp and likely resulted in a mechanical tear in the dura with flexion over time. After failing conservative treatment including hydration and caffeine, she underwent a 10 ml volume autologous epidural blood patch (EBP) under fluoroscopy

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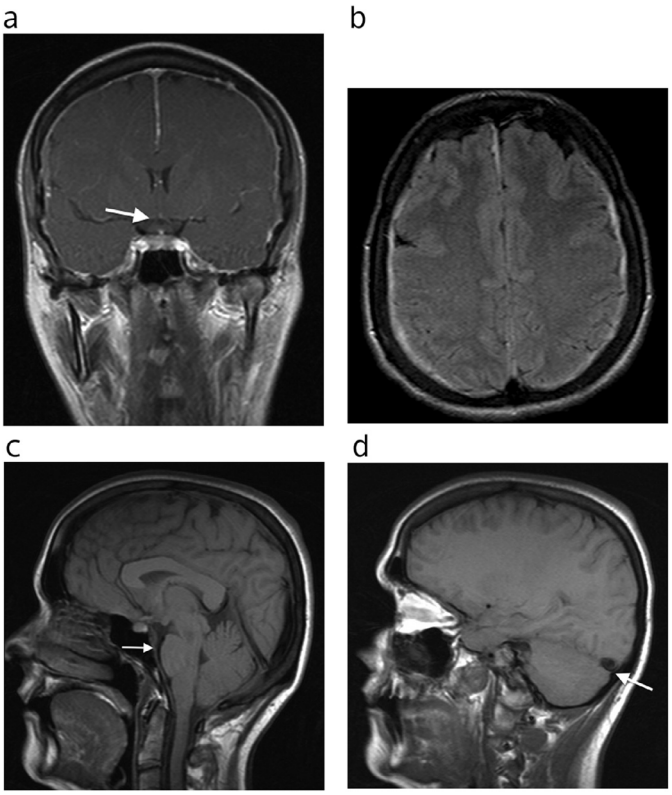

Fig 1. 41 year old female with intracranial hypotension. (A) Coronal T1 post contrast image demonstrates thin smooth pachymeningeal enhancement along the convexities and falx cerebri. Downward displacement of the brain results in effacement of the suprasellar cistern (arrow). (B) Axial FLAIR image demonstrates hyperintense signal along the convexities and falx corresponding to regions of pachymeningeal enhancement. A subdural hematoma is not present. (C) Sagittal T1 image demonstrates prominence of the dura along the prepontine cistern (arrow) and downward displacement of the cerebellar tonsils resulting in ventral displacement of the pons. (D) Sagittal T1 image demonstrates convex inferior margin of the dominant transverse sinus described as the venous distension sign (arrow).

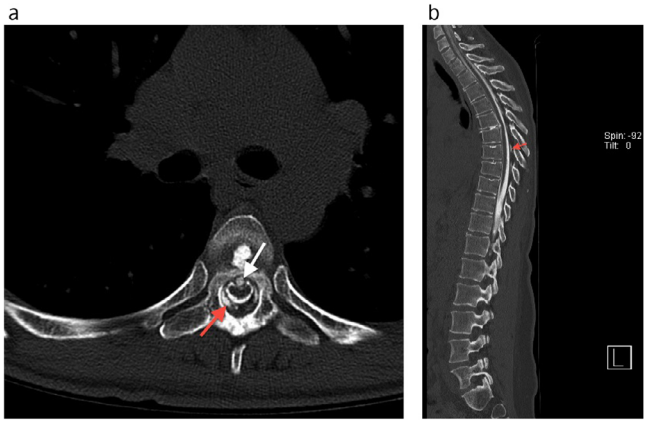

Fig. 2. 41 year old female with intracranial hypotension. Axial (A) and sagittal (B) reformatted CT myelogram images demonstrate a central calcified disc herniation (white arrow) indenting the ventral aspect of the thecal sac and distorting the thoracic cord. Extra-dural contrast material can be seen dorsally and laterally (red arrows). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

at T5-T6 which resulted in resolution of symptoms prior to hospital discharge. She underwent a second 10 ml volume autologous EBP at T5-T6 three weeks later in the outpatient setting secondary to relapse of headaches. Her symptoms have not recurred in the last 9 years.

Table 1
Imaging findings of intracranial hypotension on MRI of the brain.

Pachymeningeal enhancement
Subdural collections
Engorgement of the dural sinuses (venous distension sign)
Downward displacement of the cerebellum resulting in:
• Tonsillar descent into foramen magnum (mimicking chiari 1 malformation)
• Effacement of the suprasellar, prepontine, and prechiasmatic cisterns
• Flattening or tenting of the optic chiasm
Enlargement of the pituitary mimicking macroadenoma
Decreased size of ventricles

Table 2
Imaging findings of intracranial hypotension on MRI of the spine.

Epidural or subdural collections
Spinal dural enhancement
Engorgement of epidural venous plexus
Prominent spinal cord veins
Prominent disk or osteophyte as cause of CSF leakage

2.2. Imaging findings

The classic presentation of intracranial hypotension is orthostatic headaches, low CSF opening pressure, and smooth pachymeningeal enhancement [1,2]. Other brain MRI findings include downward displacement of the cerebellum resulting in effacement of the prepontine and prechiasmatic cisterns, flattening or tenting of the optic chiasm, subdural effusions, enlargement of the pituitary, and engorgement of the dural sinuses (Table 1) [2–4]. In some cases, there can be irregular nodular thickening of the meninges which could mimic an infectious etiology such as tuberculous meningitis. However, imaging findings such as sagging of the brainstem and engorgement of dural sinuses combined with clinical findings such as absence of fever can be used to exclude infectious pathology.

MRI findings in the spine (Fig. 3) include epidural fluid collection, dilation of the epidural venous plexus, and prominent spinal cord veins (Table 2) [5]. In rare cases, the CSF tear in the dura may be tamponaded by a herniated cord, a phenomenon referred to as idiopathic thoracic spinal cord herniation [6]. In such instances, there is regression of postural headaches and development of progressing myelopathy [6]. When imaging and clinical findings are suggestive of intracranial hypotension, examination of the entire neural axis should be performed to identify the site of CSF leakage.

CT myelography with water soluble contrast is the study of choice for detection of spinal CSF leak [1]. In addition to confirming the presence and location of CSF leak by demonstrating extra-thecal extravasation of contrast, structural abnormalities such as disc herniations, osteophytes, and meningeal diverticula as the cause of dural tear can be identified. Imaging can also reveal additional findings such as disc herniation causing spinal canal stenosis or intramedullary disease such as syringomyelia. Immediate CT after intrathecal injection or careful evaluation under digital subtraction angiography can identify the site of leakage in patients with rapid CSF leak [7]. Valsalva maneuver, walking several minutes after intrathecal injection, delayed imaging up to 4 h, and infusion of artificial CSF can increase sensitivity of finding the leak site in patients with slow flow CSF leaks [3,7,8]. Nuclear cisternography has also been used to detect the presence of a CSF leak; however it often fails to confirm the exact site of leakage [2].

3. Discussion

In a recent review of the literature, we found an additional 19 cases of CSF leak secondary to spinal pathology (Table 3) [9–22]. The mean age was 42.4 years (range: 25–57) and female to male