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Case Report

A case of spontaneous intracranial hypotension in a 45-year-old male with headache, behavior changes and altered mental status

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ABSTRACT

Spontaneous intracranial hypotension is a rare disease that results from low cerebrospinal fluid (CSF) volume caused by leakage of CSF from the spine in the absence of lumbar puncture, spine surgery, or intervention. The most common presentation is the headache that is usually but not invariably orthostatic. The underlying pathology is a CSF leak resulting from dural weakness involving the nerve root sleeves, ventral dural tears associated with calcified disc herniations, or CSF venous fistula. In severe cases, neuropsychiatric symptoms and changes in mental status may develop. Some case reports also mention gait disturbances, slurred speech, and urinary incontinence. The constellation of neuropsychiatric symptoms similar to behavior variant frontotemporal dementia in the presence of “brain sag” on MRI is known as frontotemporal brain sagging syndrome, first described by Wicklund et al. (4). The disease presents a diagnostic challenge to the primary care physicians, who are the first to see these patients. Brain and spine imaging is key to diagnoses but requires a high index of suspicion, as very rarely are all classic findings of intracranial hypotension present in the same patient. Here we discuss a case of spontaneous intracranial hypotension in a 45-year-old male patient who presented with headache, drowsiness, incoherent speech, behavior symptoms, and altered mental status.

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Introduction

Georges Schaltenbrand first described spontaneous intracranial hypotension in 1938, a German neurologist famous for his work on the motor system, cerebrospinal fluid (CSF), and multiple sclerosis [10]. Although the term implies disorder of

low CSF pressure, many recent case studies suggest that normal CSF pressure is present in some cases [11]. The underlying cause is usually low CSF volume resulting from spontaneous leakage of CSF from the spine in the absence of any spinal intervention. Although trauma is not a direct cause, many prior case reports show association with trivial trauma. The presenting symptom is often an orthostatic headache

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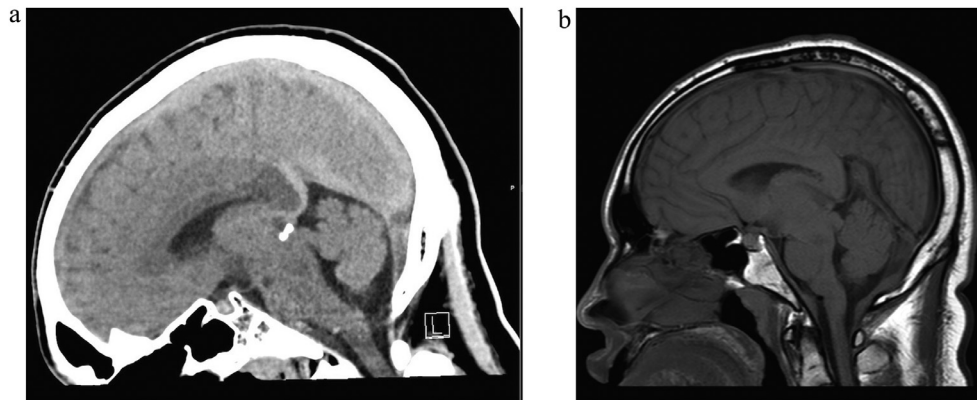


Fig. 1 – (A) Sagittal CT head depicts central brain descent, slumping of the midbrain and downward displacement of the pons. (B) Sagittal T1WI demonstrates the same findings in greater detail. There is sagging of the midbrain, downward displacement of pons, prominent pituitary and retroclival subdural hemorrhage.

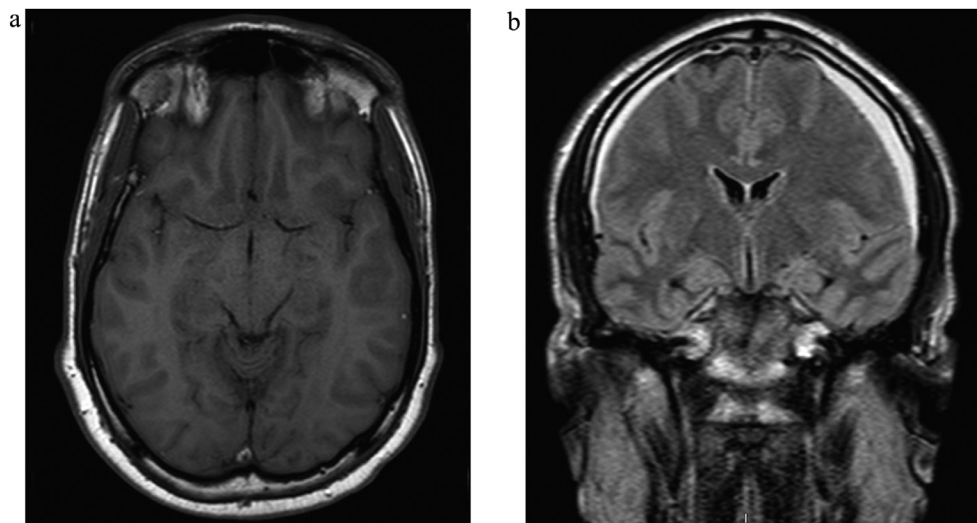


Fig. 2 – (A) Axial T1WI depicts near complete effacement of the suprasellar cistern, ambient cisterns and partial effacement of the quadrigeminal plate cistern. There is bilateral uncus herniation. (B) Coronal FLAIR depicts hyperintense bilateral frontal subdural hematomas and uncus herniation.

that occurs or worsens upon standing and improves or completely disappears when lying flat. There are multiple associated symptoms, including nausea, vomiting, photophobia, diplopia, changes in hearing, and vertigo. The diagnosis of spontaneous intracranial hypotension poses a challenge due to the variable clinical presentation and overlap with more common neurological diseases such as migraine, meningitis, and subarachnoid hemorrhage [3]. Brain and spine imaging, primarily MRI and myelography, play a vital role in diagnosing the condition.

Case report

A 45-year-old male with no past medical history came in for intractable headaches. The headaches started abruptly 1 week after sustaining a head injury with a heavy box at work. The

headache was bifrontal, throbbing, and improved on lying flat. The headache was associated with nausea and photophobia. He had multiple emergency room visits and his symptoms were attributed to migraine. He then developed a very severe headache episode while driving his mail truck, where he had to stop and lay down flat on the floor. He was brought to the emergency room and a computed tomography (CT) head and brain magnetic resonance imaging (MRI) showed bilateral frontoparietal subdural hematomas, sulcal effacement, effacement of ventricles, basal cisterns, and central transtentorial herniation. The size of the subdural hematomas was disproportionate to the mass effect and herniation. The patient underwent craniotomy for evacuation of the subdural collections. The headache improved transiently following the surgery. Six days following surgery, he developed worsening headache, agitation, memory loss, somnolence, and episodes of screaming at night and was brought to the emergency room again by his mother. He was lethargic, unable to follow com-

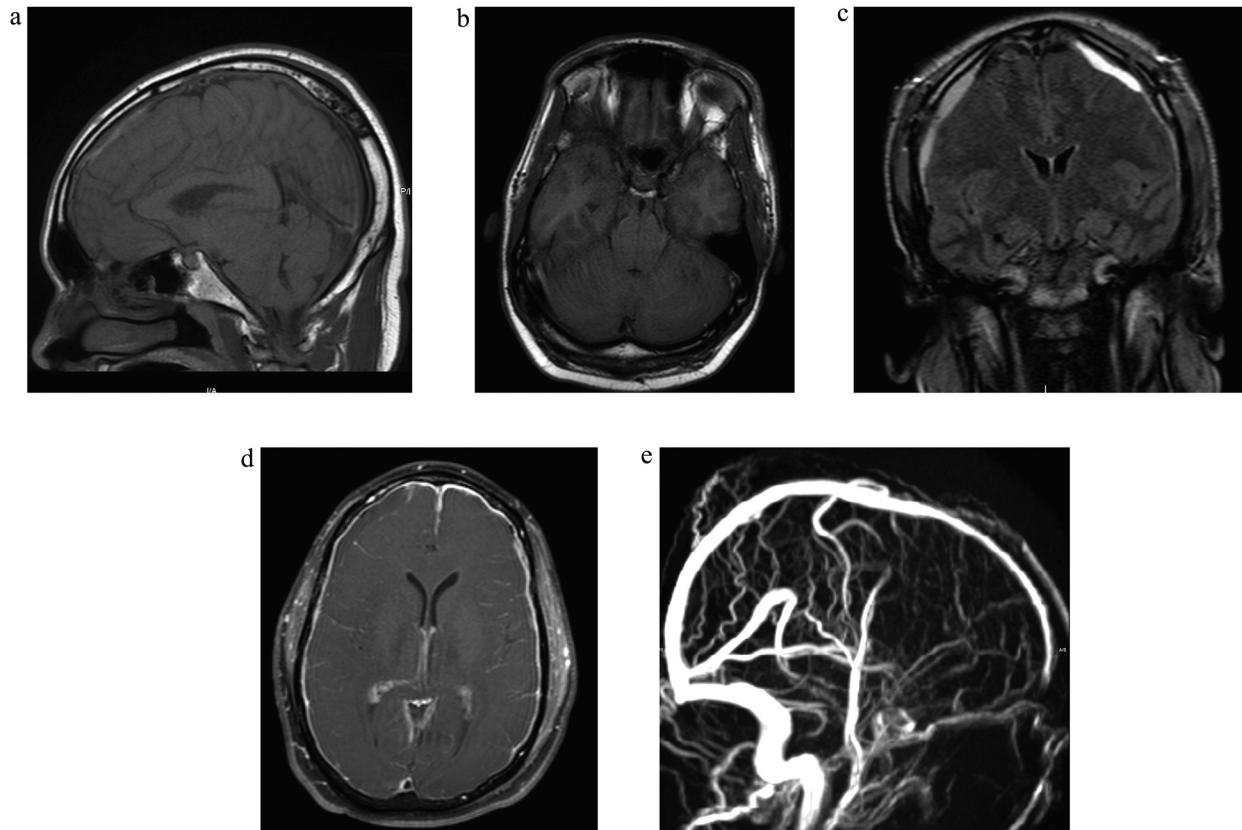


Fig. 3 – (A) Sagittal T1WI acquired 6 days following bilateral craniotomies to drain the subdural hematomas depicts exacerbation of the central brain descent with slumping of the midbrain, downward displacement of pons and cerebellar tonsils, loss of pons-midbrain angle. The pituitary is prominent and there is downward displacement of the optic chiasm. (B) Axial T1WI acquired post-operatively depicts bilateral uncal herniation and complete effacement of the suprasellar and quadrigeminal plate cistern. (C) Coronal FLAIR image acquired post-operatively depicts decrease in size of the left subdural hematoma and slight increase in the right subdural hematoma. (D) Axial post contrast T1WI depicts bilateral, diffuse and smooth pachymeningeal enhancement. (E) MRV of the brain depicts prominence of the dural venous sinuses, cortical and deep cerebral veins in the absence of occlusion or stenosis.

mands and oriented to person and place but not time. Basic laboratory workup showed no significant abnormality and a negative toxicology screen. CT head without contrast showed interval increase in the size of the right subdural hematoma, worsening effacement of basal cisterns and ventricles and a 3 mm right to left midline shift.

Brain MRI with and without intravenous gadolinium showed increasing transverse compression on the mid-brain, kinking of the brainstem, progression of central transtentorial herniation, prominent pituitary, and downward displacement of the optic chiasm. There was diffuse thickening and enhancement of the pachymeninges. The MRV showed prominence of the dural venous sinuses and cortical and deep cerebral veins without evidence of occlusion or stenosis. The MRI spine survey revealed ventral and dorsolateral epidural collections in the cervical and thoracic spine. CT myelogram was performed to look for the site of CSF leak in the spine. The CT myelogram showed ventral and dorsal epidural collections in the cervical and thoracic spine. A single point of communication between the thecal sac and epidural collection was not seen. There was an extension into the neural foramina bilat-

erally at multiple levels with extravasation of thecal contrast into the paraspinal tissues, extra-pleural space, and retrocaval soft tissues. A calcified disc protrusion was also noted indenting the ventral thecal sac and abutting the spinal cord at the T3-T4 level.

The patient was placed in Trendelenburg position, treated with intravenous hydration, and transferred to a tertiary hospital for further care. He had a lumbar epidural blood patch and T3-T4 laminectomy to repair the ventral dural defect. Postoperatively his headache improved, but his mental status kept fluctuating. He continued to have disinhibition, paranoia, memory loss, dysarthria, and inability to follow commands. He was given Seroquel to manage behavioral symptoms and dexamethasone to reduce cerebral edema. He was admitted to the Inpatient Rehabilitation unit in the hospital. An EEG showed background slowing consistent with mild diffuse cerebral dysfunction. He had 15 hours of physical and occupational therapy per week. The Seroquel was switched to Valproate to reduce drowsiness. He showed improvement in arousal and cognitive status, although he still had reduced attention span, slurred speech, tangential conversation, and re-

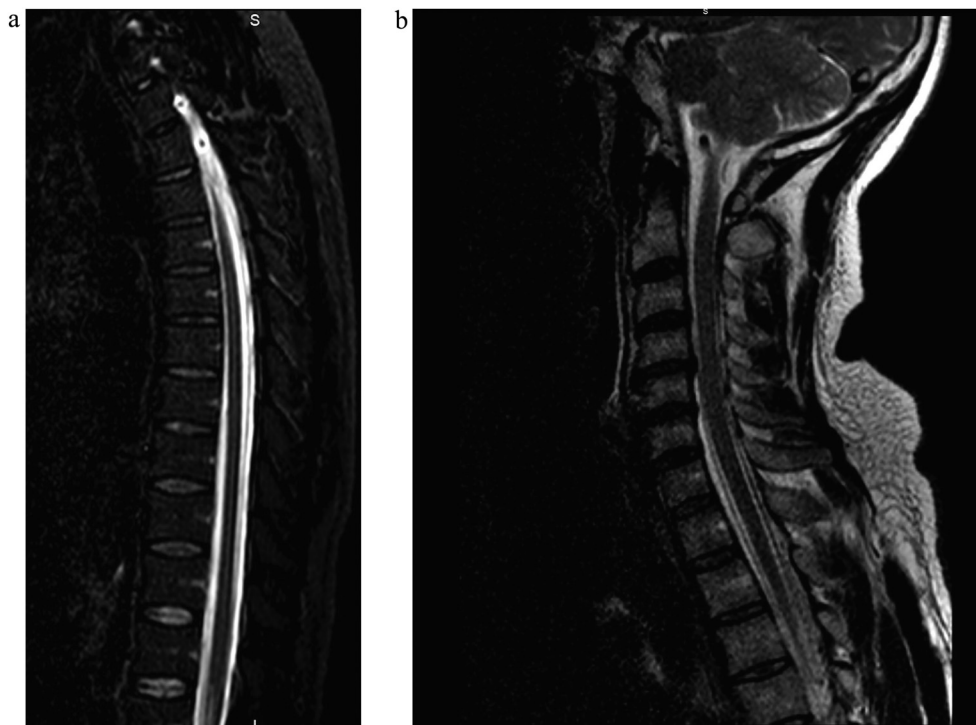


Fig. 4 – (A) Sagittal STIR image of the thoracic spine depicts dorsal epidural collection spanning the thoracic spine. (B) Sagittal T2WI of the cervical spine depicts ventral epidural collection spanning from C6 to T2 and dorsal epidural collection extending from the inferior endplate of C6 to T4.

quired frequent redirection to tasks. He was discharged under the care of his girlfriend and outpatient follow-up. He had improvements in his memory, cognition, dysarthria, and headaches. Repeat CT scan of the head two months after the T3-T4 laminectomy showed complete resolution of the subdural hematomas. The ventricles and basal cisterns were restored, and the brainstem appeared normal.

Discussion

Intracranial hypotension is a rare entity that is often difficult to diagnose; with estimated incidence of approximately 5 per 100,000 per year [5]. It is most often found in young to middle-aged adults, with female predominance [6]. The most common presenting symptom is a postural headache. Patients with intracranial hypotension most often present with a headache that changes with position, as did this patient; the headache worsens when in the upright position and is usually relieved by lying down [7]. Intracranial hypotension can be divided into 2 entities: spontaneous vs secondary. Secondary intracranial hypotension is usually iatrogenic or traumatic, often caused by procedures, such as lumbar puncture or placement of VP shunts, that may cause injury to the dura mater resulting in CSF leak [5].

On the other hand, spontaneous intracranial hypotension results from trivial trauma and weakness in the dural sac from dural tears caused by degenerative disease of the spine or

spontaneous dural dehiscence [1]. With dural injury and resultant CSF leak, there is a decrease in CSF volume, diagnosed by opening pressure of less than 60 mm of water. The hypothesis is that the postural headache results from the downward displacement of the brain stemming from a decrease in CSF volume with subsequent traction on pain-sensitive structures [3]. “Sagging of the brain” from loss of buoyant force of CSF also leads to traction on cerebral and cerebellar veins, meninges, and cervical nerves, causing many other associated symptoms in addition to headaches, such as nausea, vomiting, photophobia, cranial nerve palsies, and many others [8]. Severe brain sagging may result in a coma from diencephalic or hindbrain herniation [9].

Due to the rarity of the entity, the various divergent presentations, and the numerous other associated symptoms, spontaneous intracranial hypotension is a difficult diagnosis to make. For example, there have been written case reports concerning patients with CSF leaks with either normal opening pressure, absent pachymeningeal enhancement, no orthostatic headache, or CT myelography showing only meningeal diverticula instead of an actual leak [4]. Additionally, many of the associated symptoms may mimic other pathologies. For example, commonly seen signs and symptoms, such as nausea, vomiting, photophobia, and posterior neck pain/stiffness may mimic subarachnoid hemorrhage or infectious meningitis. Due to low CSF volume, traumatic tap is not uncommon and frequently leads to confusion with subarachnoid hemorrhage. High protein content and pleocytosis in CSF analysis can mislead physicians to diagnose meningitis. Cerebellar

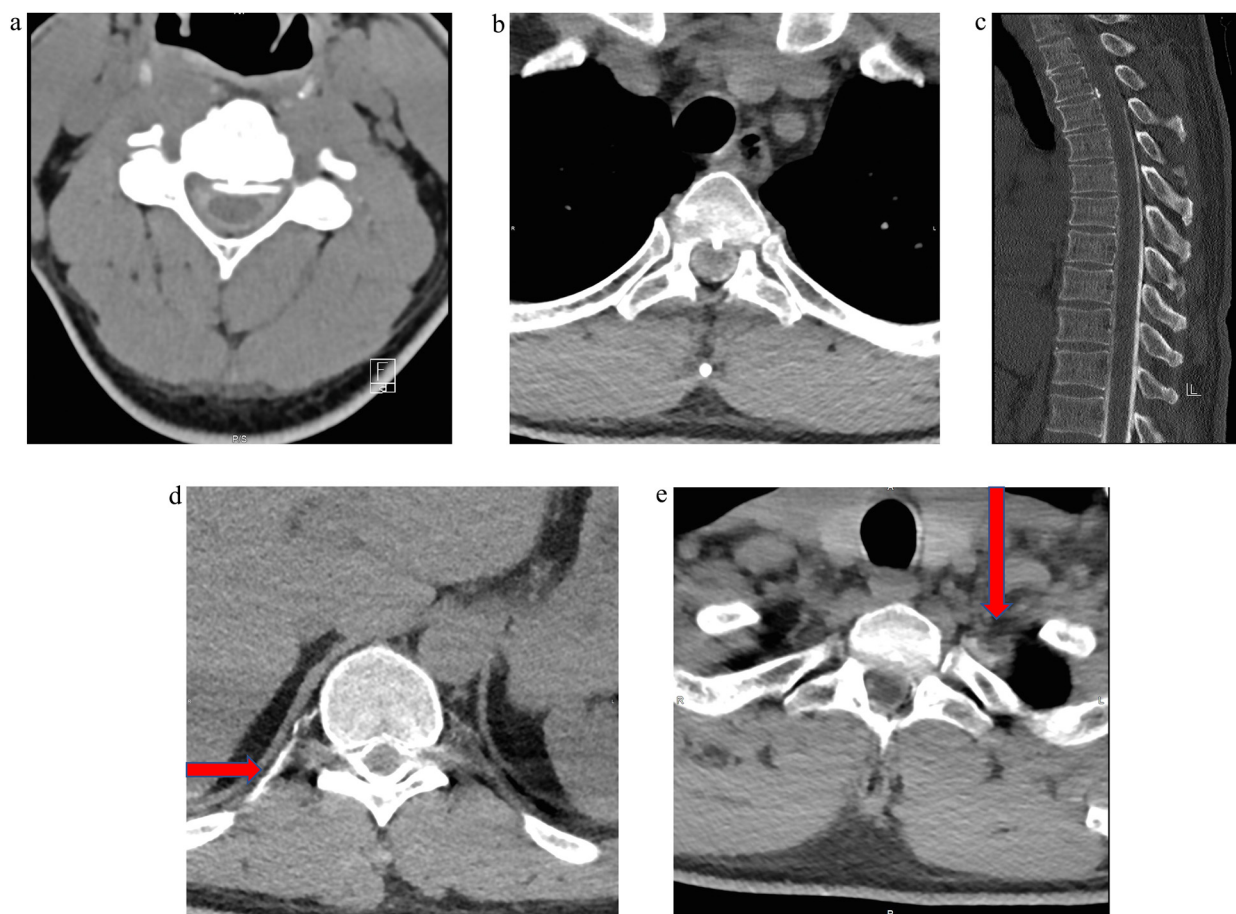


Fig. 5 – (A) Axial CT image of the cervical spine post myelography depicts hyperdense ventral epidural collection. Note that the collection is hyperdense compared to the intrathecal contrast due to pooling of contrast in a confined space. (B) Axial CT image of the thoracic spine depicts a calcified disc protrusion at T3-T4 which indents the thecal sac and abuts the ventral spinal cord. (C) Sagittal CT of the thoracic spine depicts a solitary calcified disc protrusion at T3-T4. (D) Axial CT image of the thoracic spine post myelography depicts extravasation of contrast into the right retrocrural soft tissues at T10-T11. (E) Axial CT image of the thoracic spine post myelography depicts contrast extending into the extra-pleural space at the left lung apex.

tonsillar herniation can also be mistaken for Chiari I malformation; some patients have undergone decompressive posterior fossa surgery with no relief [3]. In one of the studies, only 1 out of 18 patients were diagnosed with spontaneous intracranial hypotension at first physician encounter; the remaining 17 patients had to visit a range of 1-6 physicians. A total of 33 working diagnoses were designated, and the time to diagnosis had a mean of 13 months and a median of 5 weeks. [3].

Our patient also presented with psychiatric instability and behavioral changes compared to baseline with progressive decline in executive functions, preventing him from working. Frontotemporal brain sagging syndrome, defined as the constellation of clinical presentation like that of behavioral variant frontotemporal dementia and radiologic findings of inferior brain herniation, has been described in past studies on patients with intracranial hypotension [4]. One participant from the study showed similar symptoms as our patient, with orthostatic headache, imaging findings of intracranial hypotension, and behavioral disturbances [4]. Behavior variant fron-

totemporal dementia (bvFTD) is a subtype of frontotemporal dementia characterized by disinhibition, apathy or inertia, diminished social interest, compulsive behavior, hyperorality, and dietary changes, deficits in executive functioning, and relative sparing of episodic memory and visuospatial functioning [12]. The patients with FBSS do not have all the typical symptoms of bvFTD. The mimicking features include disinhibition, the age at onset, and male predominance. FBSS is usually characterized by daytime somnolence and dysarthria, as was seen in our patient, and dysphagia, gait disturbance, ocular abnormalities, or movement disorders. On imaging, the bvFTD shows atrophy in the frontal and temporal lobes, which is not seen in FBSS; however, FBSS indeed shows frontotemporal hypometabolism on PET and SPECT like bvFTD [4]. The frontal lobes have widespread connections to different brain regions, including the brainstem and cerebellum, known as frontal network systems. These systems play a crucial role in cognition, including working memory, attention, initiation, inhibition, monitoring, emotion, and language [16]. The two

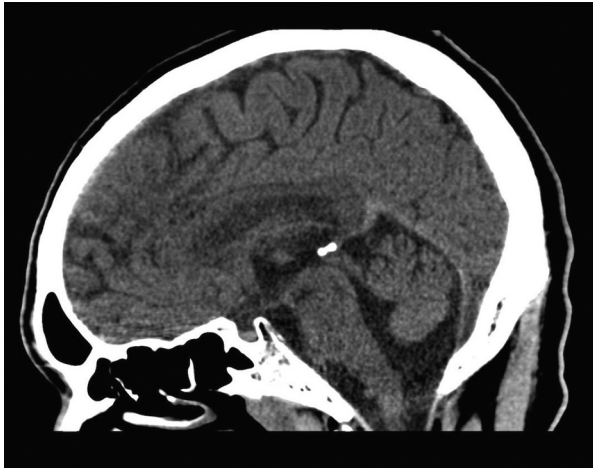


Fig. 6 – Sagittal CT head 2 months after repair of dural defect shows normal appearing brainstem with restoration of the pons-midbrain angle, the prepontine and suprasellar cisterns and normal sized pituitary.

most critical frontal networks are the salience and default mode networks. The salience network consists of the anterior cingulate and frontoinsular cortices, hypothalamus, thalamus, amygdala, ventral striatum, and several brainstem nuclei [14]. The default mode network consists of the posterior cingulate cortex, precuneus, medial prefrontal, and inferior parietal cortices [15]. The central herniation and brainstem distortion lead to mechanical forces on the frontal network systems, leading to disturbances analogous to the network-topathies of bvFTD [4]. Somnolence may result from dysfunction of the midbrain reticular formation [13].

Ranging from highly variable, nonspecific neurologic symptoms to association with psychiatric manifestations and with many potential mimickers and pitfalls, spontaneous intracranial hypotension is a difficult diagnosis to make. For these reasons, there must be a high index of suspicion when there are even a few of the clinical symptoms and imaging features of spontaneous intracranial hypotension. Multiple imaging features may be present in patients with spontaneous intracranial hypotension. One of the most frequently encountered findings on MRI is a fluid collection that often extends over 5 or more spinal segments [2]. Our patient's MRI displayed both dorsal epidural collections spanning the entire thoracic spine and ventral epidural collection spanning from the inferior endplate of C6 to the inferior endplate of T2. More common image findings include pachymeningeal enhancement and inferior displacement of the brain, brainstem, and cerebellum, signified by effacement of the suprasellar cistern, bowing of the optic chiasm over the pituitary fossa, flattening of the pons against the clivus, and downward displacement of the cerebellar tonsils [3]. Our patient demonstrated pachymeningeal enhancement and inferior displacement of the brain evidenced by effacement of multiple basal cisterns, bilateral uncal herniation, slumping of the midbrain, and inferior displacement of the pons. The patient also presented with concurrent, bilateral subdural hematoma along the frontoparietal convexity, masking the source of mass effect. Other

MRI findings that can be found in the spine include dilated epidural veins, engorgement of the epidural venous plexus, and dural enhancement of the spine [2] (Figs. 1-6).

Patient consent

Verbal consent was obtained from the patient's caregiver.

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