Spontaneous intracranial hypotension

- Targeted Versus NonTargeted Epidural Blood Patch for Spontaneous Intracranial Hypotension: A Systematic Review and Meta-Analysis
- Outcomes of CT-Guided Targeted Epidural Patching For Lateral Dural Tears In Spontaneous Intracranial Hypotension: A Multicenter Retrospective Cohort Study
- Simulation of cerebrospinal fluid (CSF)-venous fistula embolization in a swine model: A technical video
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- Transdural Approach for Repair of a Spontaneous Ventral Thoracic Cerebrospinal Fluid Leak Using Muscle Plug: 2-Dimensional Operative Video
- Symptomatic cerebral spinal fluid leak (or intracranial hypotension) due to sacral extradural cyst rupture after sacral fracture: illustrative case
- Intradural venous engorgement of CSF-venous fistula mimics spinal dural arteriovenous fistula on MRI: A novel case report and review of literature

Spontaneous intracranial hypotension (SIH) is a syndrome characterized by cerebrospinal fluid hypovolemia and low cerebrospinal fluid pressure related to cerebrospinal fluid fistula, that results in various symptoms.

Intracranial hypotension is a condition in which there is negative pressure within the brain cavity.

From an evolutionary standpoint, as we became bipedal, certain accommodating mechanisms developed to prevent during upright positioning the overdrainage of fluid (CSF or blood) from the cranial to the spinal compartment, which could lead to intracranial hypotension.

Epidemiology

Spontaneous intracranial hypotension (SIH) is caused by spinal cerebrospinal fluid (CSF) leaks, which result in continued loss of CSF volume and multiple debilitating clinical manifestations. The estimated annual incidence of SIH is 5/100,000. Spontaneous intracranial hypotension (SIH), once believed to be rare, is now more commonly recognized.

Etiology

see Spontaneous intracranial hypotension

Cerebrospinal fluid leak from the spinal canal:

A leak following a lumbar puncture (spinal tap).

A defect in the dura

Sometimes following exertion such as swinging a golf club.

A congenital weakness.

Following spinal surgery.

Following spinal trauma.

Following a shunt procedure for hydrocephalus.

Lumboperitoneal shunt.

Ventriculoperitoneal shunt with a low pressure valve.

In some cases, spinal CSF leaks can lead to a descent of the cerebellar tonsils into the spinal canal, similar to a Chiari malformation.

Large spinal dural defects can lead to herniation of the spinal cord into the defect.

Pathophysiology

see Spontaneous intracranial hypotension pathophysiology.

Clinical features

Intracranial hypotension clinical features.

Treatment

see Intracranial hypotension treatment.

Complications

Intracranial Hypotension complications.

Outcome

If the cause of the intracranial hypotension can be identified, the outcome following treatment is typically excellent.

Etiology

Cerebrospinal Fluid-Venous Fistula are increasingly identified as a cause of spontaneous intracranial hypotension (SIH).

Practically all cases of spontaneous intracranial hypotension results from spontaneous cerebrospinal fluid leaks, often at the level of the spine and only rarely from the skull base.

The triad of orthostatic headaches, diffuse pachymeningeal enhancement on head imaging and low CSF opening pressure is considered the hallmark of these leaks [1)].

German neurologist Schaltenbrand reported that orthostatic headache by low cerebrospinal fluid pressure in 1938. This disease came to be known after development of radiological diagnosis in 1990.

Shinonaga et al reported that cerebrospinal fluid leak is induced in the whiplash sequelae after traffic accident in 2003. Cerebrospinal fluid hypovolemia got into the news social. A lot of doctors deny the cerebrospinal fluid leak after mild traffic accident. The Cerebrospinal Fluid Hypovolemia Society is started up in 2003 and 11 research meeting held until today. The research group of Ministry of Health, Labour and Welfare was made in 2007. The image diagnostic criteria of cerebrospinal fluid leakage syndrome model were made in 2012. Neither the mechanism of the cerebrospinal fluid leak nor the mechanism of symptoms are understood well. The pathophysiology of cerebrospinal fluid hypovolemia is expected by researching the cerebrospinal fluid circulation [2)]

Shinonaga et al. (2001) suggested that more than 80% of patients with whiplash injury complaining vertigo and dizziness showed cerebrospinal hypovolemia on radioisotope cisternography (111In-DTPA). However, neuro-otological studies to investigate the pathophysiological mechanisms underlying these symptoms have been insufficient. In a study, patients complaining of these symptoms with CSF hypovolemia after traffic accidents were investigated with posturography and electronystagmography (ENG). Fourteen patients (4 men, 10 women; 24-52 yr) were examined with posturography and showed parameters (tracking distance & area) significantly ($p<0.01$) larger than those of healthy subjects. Among them, five cases (1 man, 4 women; 31-52 yr) were further investigated with ENG. The slow phase peak velocities of optokinetic nystagmus (OKN) and optokinetic-after nystagmus (OKAN) were significantly ($p<0.01$) reduced (62.64±6.9 SD deg/sec, 60.76±10.74 SD deg/sec, respectively) and frequencies of OKN were reduced (139.7±10.75 SD), while the ocular smooth pursuit was relatively preserved. Magnetic resonance images (sagittal view) of these five patients demonstrated the downward displacement of the cerebellar tonsils and flattening of the pons, which are characteristic features of CSF hypovolemia, called “brain sagging.” The results suggest that brain sagging due to CSF hypovolemia impairs vestibular and vestibulocerebellar functions, which may cause dizziness and vertigo [3)].

As a cause of acute clinical deterioration after aneurysmal clipping, CSF hypovolemia is likely under-recognized, and may actually be misdiagnosed as vasospasm or brain swelling. We should always take the etiology of CSF hypovolemia into consideration, and especially pay attention in patients with pneumocephalus and subdural fluid collection alongside brain sag on computed tomography. These patients are at higher risk developing of pressure gradients between their cranial and spinal compartments, and therefore, brain sagging after Lumbar drainage (LD), than after ventricular

drainage. We should be vigilant to strictly manage LD so as not to produce high pressure gradient [4)].

Pathophysiology

Spontaneous intracranial hypotension pathophysiology.

Clinical features

Intracranial hypotension clinical features

Diagnosis

see Spontaneous Intracranial hypotension diagnosis

Complications

see Intracranial hypotension complications.

Treatment

Spontaneous intracranial hypotension treatment.

Case series

Patients with spontaneous intracranial hypotension diagnosed with a Cerebrospinal Fluid-Venous Fistula between January 2021 and December 2022 in which the area of CVF(s) was covered by both diagnostic modalities were included. LD-CTM immediately followed LD-DSM without repositioning the spinal needle, and the second half of the contrast agent was injected at the CT scanner. Patients were awake or mildly sedated. Retrospectively, two neuroradiologists evaluated data independently and blinded for the presence of CVF.

Twenty patients underwent a total of 27 combined LD-DSM/LD-CTM examinations (4/20 with follow-up and 3/20 with bilateral examinations). Both raters identified significantly more CVFs with LD-CTM than with LD-DSM (rater 1: 39 vs 9, P<0.001; rater 2: 42 vs 12, P<0.001). Inter-rater agreement was substantial for LD-DSM (κ=0.732) and LD-CTM (κ=0.655). The results remained significant after considering the senior rating for cases of disagreement (39 vs 10; P<0.001), and no CVF detected on LD-DSM was missed on LD-CTM.

In this study, Lateral decubitus CT myelography has a higher diagnostic yield for the detection of CVFs

than LD-DSM and should supplement LD-DSM, but further studies are needed. LD-CTM can be easily acquired in awake or mildly sedated patients with the second half of contrast injected just before CT scanning, or it may be considered as a stand-alone investigation [5].

The study provides valuable insights into the diagnostic yield of LD-DSM and LD-CTM for CVF detection in SIH patients. However, it has limitations related to sample size, study design, and the need for further validation. The findings support the potential role of LD-CTM as a more sensitive diagnostic tool, but its clinical implications and safety considerations should be explored in more detail.

Ultrahigh-Resolution Cone-Beam Computed Tomography [6]

Direct intraoperative visualization of CVF using intrathecal fluorescein. CVF can be identified intraoperatively using fluorescein dye, which can be a valuable adjunct for the surgeon confronted with this disease [7].

Tanaka et al retrospectively studied 40 consecutive patients (female:male = 28:12, median age 41.5 years) treated under clinical diagnoses of SIH satisfying the International Classification of Headache Disorders 3rd edition criteria, including 37 patients (92.5%) with diffuse pachymeningeal enhancement. The patients were divided into two groups by age and sex, and the clinical and neuroimaging findings in each group were investigated.

Acute onset (female:male = 82.1%:50.0%, P = .042), severe headache (75.0%:41.7%, P = .045) occurred with higher frequency in females than in males, and SDH occurred with lower frequency in females than in males (28.6%:75.0%, P = .006). Duration until the consultation (2:14 days, P = .022), SDH thickness (0:7.1 mm, P = .001), and iter displacement (1.6:7.1 mm, P = .004) was greater in males. Acute onset (Younger [≤40 years]: older [>40 years] = 94.1%:56.5%, P = .012), occurred with higher frequency in younger patients, and duration until the consultation (1:5 days, P = .001), frequency of SDH (17.7%:60.9%, P = .010), SDH thickness (0:5.9 mm, P = .003), in older patients. All nine patients with thunderclap headache were female, with median age of 37 years.

More severe clinical symptoms with acute onset were observed in females and younger patients of SIH. Comparatively rare subdural hygroma/subdural hematoma on magnetic resonance imaging might result from the shorter duration to diagnosis in females and younger patients [8].

Case reports

2016

A 43-year-old male complained of experiencing orthostatic headaches for 2 months without neurological signs. The patient worsened in a local hospital and was transferred to the Sir Run Run

Hospital. Brain computed tomography showed bilateral SDH with a midline shift. The patient underwent emergency trephination in the left frontal temporal region. Postoperative magnetic resonance myelography showed a CSF leak originating at the T11-L2 level. As a consequence of clinical deterioration of the patient, EBP was subsequently performed at the T12-L1 level. The headache was rapidly relieved and later the SDH was completely absorbed. This case report and literature review aims to remind clinicians that SIH can cause SDH and that EBP is a viable treatment option [9)].

A 59-year-old man presented with a case of SIH that manifested as a bilateral chronic SDH. He developed fatal extensive pneumocephalus and SDH re-accumulation as a complication of burr-hole drainage. Despite application of an epidural blood patch, the spinal cerebrospinal fluid leak continued, which required open spinal surgery. Chronic SDH management should not be overlooked, especially if the exact cause has not been determined. When chronic SDH assumed to be associated with SIH, the neurosurgeon should determine the exact cause of SIH in order to effectively correct the cause [10)].

2015

A 58-year-old woman with an altered mental status who had visited a local hospital and in whom a brain CT showed a unilateral subdural hematoma with a marked midline shift. She was referred to the Department of Neurosurgery, Fujita Health University Hospital, Toyoake, Japan because of her neurologic deterioration after hematoma evacuation. A CT myelography revealed a massive CSF leakage in the entire thoracic epidural space. She made a full neurologic recovery following blood patch therapy. Our case is unique and educational because the suspicion for SIH as an underlying cause of subdural hematoma is warranted in nongeriatric patients not only with bilateral but also unilateral lesions. An immediate search for CSF leakage may be important in cases with failed hematoma evacuation surgery [11)].

2014

A 34-year-old man presented with acute postural headache. The first cerebral computed tomography scan was normal. Lumbar puncture showed hyperproteinorachy at 2 g/L with six lymphocytic cells. The headache became very intense. At admission, clinical examination was normal. Ophthalmological examination did not show any abnormalities. Encephalic magnetic resonance imaging (MRI) showed bilateral subdural hematoma with tonsillar descent simulating Chiari type I malformation. After surgical drainage and symptomatic treatment, the patient was discharged with no recurrence.

Spontaneous intracranial hypotension is associated with simple clinical presentation, orthostatic headache, and characteristic MRI findings. Misdiagnosed, it leads to unnecessary procedures [12)]

A 23-year-old woman was injured in a rear-end collision. She had general malaise and posterior neck pain, which were more severe when she was in an upright position. Magnetic resonance imaging (MRI) revealed the presence of cerebellar tonsil descensus and syringomyelia in the spinal cord. Radioisotope (RI) cisternography showed signs of an early accumulation of RI in the bladder, and a

delayed accumulation of RI in the cerebral fornix.

Hatae et al considered the possibilities of cerebrospinal fluid (CSF) hypovolemia and congenital Chiari type-1 malformation as being responsible for her headache. To obtain a definitive diagnosis, we performed gadolinium (Gd)-enhanced MR cisternography and found evidence of CSF leakage. We performed an epidural blood patch (EBP), and her symptoms resolved. In 2 years since the episode, her symptoms have not recurred, and additional treatment has not been required. In addition, MRI performed 2 years after the EBP did not reveal any changes. There seems no previous report which described successful differentiation of pre-existing congenital Chiari type-1 malformation from the acquired one caused by symptomatic CSF hypovolemia. Because treatment protocols differ between these two conditions, the establishment of a correct diagnosis is important [13].

A 61-year-old man with thickening of the dura mater associated with the presence of subdural collections as a consequence of cerebral spinal fluid hypovolemia (CSFH) and hypertrophic pachymeningitis (HP) as presentation of systemic lupus erythematous (SLE). The patient complained about fatigue, musculoskeletal pain, headache and skin lesions. In the laboratory tests minimal normocytic anemia, mild leukopenia, polyclonal hypergammaglobulinemia and antinuclear antibodies (ANA), anti-double-stranded DNA antibodies (dsDNA), antibodies against extractable nuclear antigens (ENA) type SSA-Ro, anti-Smith antigen antibodies (anti-Sm) and anti-ribonucleoprotein antibodies (anti-RNP) were detected. Cranial magnetic resonance imaging (MRI), with and without gadolinium enhancement, revealed generalized thickening of the dura mater more severe at the right parieto-occipital lobes with the presence of subdural collections. The patient was diagnosed with SLE associated both with CSFH and HP. A conservative treatment with prednisone 60 mg daily, mycophenolate mofetil (MMF) 1 g daily and hydroxychloroquine 200 mg twice a day was started with significant clinical and radiological improvement (almost complete resolution of the subdural collections and clear decrease of meningeal thickness). The authors emphasize that HP associated with CSFH in the context of SLE is a rare entity, which makes this case unique [14]

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