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Spontaneous Intracranial Hypotension: What An Infectious Disease Physician Should Know?

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1. Introduction

Although the syndrome of spontaneous intracranial hypotension (SIH) is not an infectious disease, it is commonly involved in the differential diagnoses of meningitis and encephalitis. It is a clinical (headache, fever, even neck stiffness) and laboratory (cerebrospinal fluid (CSF) abnormalities) challenge for the physician. Considering this syndrome especially under some settings and taking care of the characteristic imaging findings would contribute to the diagnosis. We believe in that the physicians who care with these central nervous system (CNS) infections should be aware of this syndrome. Some patients under the suspicion of encephalitis have the SIH that could be diagnosed by approaches described in this chapter.

Patients presenting with fever, headache, stiff neck, nausea, vomiting and some other neurological signs suggesting meningeal irritation are always taken seriously and usually have a similar diagnostic algorithm. Differential diagnosis is usually based on the results of CSF analysis. In some cases, characteristics of the headache are the major factor determining the way to establish the diagnosis. Headache with a positional pattern, that occurs shortly after assuming an upright position and relieves by lying down, so called "orthostatic headache" is a distinctive symptom of SIH syndrome.

The spontaneous form of intracranial hypotension was first described by a German neurologist Georg Schaltenbrand in 1938 [1]. He recognized that "aliquorrhea," or as subsequently named "hypoliquorrhoa" a deficiency in cerebrospinal fluid, could result in headaches predominantly when upright. Since the introduction of magnetic resonance image (MRI) in daily diagnostic use in the early 1990s, much has been learned about SIH.

All or essentially all SIH cases are related with a spontaneous spinal CSF leak mostly at the cervicothoracic junction or along the thoracic spine [2].

SIH is an important and relatively frequent cause of newly onset daily persistent positional headaches in young and middle-aged individuals. Women are effected more frequently than men with a ratio of approximately 1.5/1. It is diagnosed about half as frequently as spontaneous subarachnoid hemorrhage and its incidence is estimated to be five per 100 000 [3].

Throbbing headache occurring or worsening in upright position and improving after lying down, so called “orthostatic headache”, low CSF pressure, and diffuse pachymeningeal enhancement on brain magnetic resonance imaging (MRI) are the major features of the classic syndrome. Many other signs and symptoms may associate.

2. Diagnostic criteria of SIH

The diagnostic criteria used to verify the SIH cases are on shown on Table 1.

The clinical presentations and radiological findings may vary. Diagnosis is largely based on clinical suspicion, cranial MR findings and myelographic detection of dural leak. The positional characteristics of the headache should always be questioned in patients admitting with meningeal irritation signs. Orthostatic headache, characteristic imaging features on MRI and instantaneous improvement of symptoms with a successful blood patch are key points of differential diagnosis from viral meningoencephalitis or other causes of aseptic meningitis syndrome.

A. Diffuse and/or dull headache that worsens within 15 minutes after sitting or standing, fulfilling criterion D and with ≥ 1 of the following:

1. Neck stiffness
 2. Tinnitus
 3. Hypacusis
 4. Photophobia
 5. Nausea
-

B. At least 1 of the following:

1. Evidence of low CSF pressure on MRI (eg, pachymeningeal enhancement)
 2. Evidence of CSF leakage on conventional myelography, CT myelography, or cisternography
 3. CSF opening pressure <60 mm H₂O in sitting position
-

C. No history of dural puncture or other cause of CSF fistula

D. Headache resolves within 72 hours after epidural blood patching

Table 1. Diagnostic Criteria for Headache Due to Spontaneous Spinal CSF Leak and Intracranial Hypotension According to the International Classification of Headache Disorders, 2nd Ed42 (4)

3. SIH and aseptic meningitis

Patients admitted with fever, headache and CSF findings revealing lymphocytic pleocytosis, elevated protein concentration and normal glucose levels are prone to be misdiagnosed as viral meningoencephalitis or usually aseptic meningitis [5].

The clinical presentation of aseptic meningitis is generally nonspecific, with fever, headache, nausea and vomiting, occasionally accompanied by photophobia and a stiff neck. Physical examination typically reveals signs of nuchal rigidity

The syndrome of “aseptic meningitis” including differing etiologies and disorders, presents a diagnostic challenge to the clinician [6]. Although many infectious and noninfectious etiologies exist for this syndrome, viruses, especially nonpolio enteroviruses, are the most common (>85%) and most important agents encountered. Although seasonal variation is relative and not absolute, enteroviruses are most likely to be the cause of aseptic meningitis occurring during the summer or fall. The onset of symptoms is characteristically abrupt and typically includes headache, fever, nausea or vomiting, malaise, photophobia, and meningismus.

Because the presenting signs and symptoms of enteroviral meningitis are not distinctive, tuberculosis meningitis, herpes simplex encephalitis, HIV encephalitis and parameningeal infection that may mimic aseptic meningitis in their initial presentations must not be overlooked.

SIH may mimic aseptic meningitis. The main features of aseptic meningitis and SIH cases are compared in a case series newly reported from Turkey [7]. Various clinical and laboratory features of 11 consecutive cases of SIH and 10 consecutive cases of aseptic meningitis are given below (Table 2).

All of the 11 patients with SIH reported that their headache was occurring or worsening within minutes or hours assuming the upright position and improving by lying down, defined as “orthostatic headache”. All met the diagnostic criteria defined by International Headache Society. The median duration of sudden-onset orthostatic headache was 10 days, ranging between 1 to 30 days. Five cases (5/11) had a previous diagnosis of migraine because of chronic headache. The newly onset orthostatic headache was throbbing and diffuse in all cases distributing from posterior neck (5/11), from frontal area (4/11), from left temporal region (1/11) and from left parietal region (1/11) to the whole cranium. The typical positional characteristics of the headache were noticed with further questioning of the patients.

Hearing changes, disturbed sense of balance, and nausea were noted in all patients. Posterior neck pain and vomiting were described in 9, tinnitus in 3 patients, and echoing in 1 case.

Stiff neck was detected in 5 patients, and fever (axillary; >37.3°C) in 7 patients. The highest temperatures of those with fever were measured as follows; 38.7 °C, 38.3 °C, 38.1 °C, 38 °C, 38.9 °C, 38.2 °C and 38.5 °C.

Eight of 11 cases had visual changes as blurring (5/11) and diplopia (5/11).

Among neurological disorders photophobia (8/11), phonophobia (8/11) were most frequent followed by subtle cognitive deficits (5/11), amnesia (4/11), confusion & syncope (3/11), dysgeusia (3/11), facial numbness (2/11), convulsion, hyperexcitability, dysarthria, ataxia, facial weakness, facial spasm (each 1/11).

| | CLINICAL SIGNS & SYMPTOMS | | | | | CSF FINDINGS (Mean±SD) | | | | | | |
|---------------------------|---------------------------|-------|-------------------|------------|---|--|--|--|--|----------------------------|--------------------|-------------------|
| | Headache | Fever | Nausea – Vomiting | Stiff Neck | Opening pressure (mmH ₂ O) (NR: 50-180)* | Leukocyte (/ mm ³) (NR:0-5)* | Lymphocyte (/ mm ³) (NR:0-5) | Protein concentration (mg/dl) (NR:15-45) *** | CSF Glucose /blood glucose (mg/dl) (NR: 40-80) | Enterovirus PCR positivity | HSV PCR positivity | TB PCR positivity |
| SIH (n=11) | 11/11 | 7/11 | 10/11 | 5/11 | 59 ±16.4 | 229±200 | 76 ± 5 | 55±49 | 61± 12/102 ±21 | 0/11 | 0/10 | 0/10 |
| Aseptic Meningitis (n=10) | 10/10 | 6/10 | 6/10 | 5/10 | 160±28.4 | 360±50 | 84±13 | 93±92 | 63±11/ 114±13 | 2/10 | 0/10 | 0/10 |

SIH: Spontaneous intracranial hypotension, CSF: Cerebrospinal fluid, NR: normal range, PCR: polymerase chain reaction, HSV: Herpes simplex virus, TB: tuberculosis

* Only five were measured.

** Pleocytosis was detected in only four SIH cases while all (n=10) cases of aseptic meningitis had lymphocytic pleocytosis varying between 32 and 1340 /mm³.

*** Four SIH and seven aseptic meningitis cases had elevated CSF protein levels.

Table 2. Comparison of SIH and Aseptic Meningitis Cases

4. SIH and encephalitis

Despite the benign character of SIH, some rare cases may present with severe neurological findings. A few cases of SIH are reported whose chief clinical manifestation were dif-

fuse severe encephalopathy with marked depression of consciousness, hyperexcitability or convulsion [7,8].

A 40 year old female [8] was admitted to Stanford University School of Medicine in California with a progressive cognitive decline of 2 to 4 weeks' duration. She developed a newly onset diffuse headache in orthostatic nature one month ago and she suffered a brief generalized seizure 2 weeks before admission. Her computerized tomography (CT) scan and electroencephalography (EEG) were normal and she was diagnosed as SIH with diffuse pachymeningitis on MRI, low CSF opening pressure (60 mm/H₂O) immediate clinical improvement responding to placement of epidural blood patch.

Two similar cases with SIH, with no defined preexisting comorbidities and newly pre-diagnosed as meningoencephalitis, were reported from Istanbul-Turkey. Both were young (29 and 21 years) males, brought to emergency departments of different hospitals in coma with a 3-year of time interval. One had a generalized tonic-clonic convulsion and the other who was evaluated as nonconvulsive status had hyperexcitability. Antiepileptic agents were administered for both before admission to the university hospital where the final diagnosis was established.

The first diagnostic steps for these cases were aimed to exclude Herpes encephalitis due to its high frequency and being a medical emergency. Although the patients were admitted with impaired conscious and convulsions, there were no signs of contrast enhancement (e.g. temporal, parietal or frontal lobe) suggesting HSV 1 involvement, the CSF opening pressures were slightly low (60 and 90 mmH₂O consequently) and HSV 1 PCR results were negative.

The diagnosis of SIH was established on the basis of specific cranial MR images, negative CSF findings and the prompt response to blood patch within 72 hours.

The most common cause of non-epidemic (not affecting a large number of people at once) encephalitis in developed countries is the herpes simplex virus. The most common signs of acute viral encephalitis are fever, headache, and a change in level of consciousness. Other common signs are the eyes becoming sensitive to light (photophobia), confusion, and sometimes seizures.

Some people exposed to insect-borne encephalitis viruses do not develop symptoms of encephalitis. They may only experience low-grade fever, drowsiness, and flu-like symptoms of malaise (general feeling of illness) and myalgia (muscle aches). Headache, vomiting, and sensitivity to light may follow. The epidemiological relatedness and a history of a travel, recall of an insect exposure are useful to exclude this type of encephalitis.

Symptoms and signs of meningeal irritation (photophobia and nuchal rigidity) are usually absent with a pure encephalitis but often accompany a meningoencephalitis. Patients with encephalitis have an altered mental status ranging from subtle deficits to complete unresponsiveness. Seizures are common with encephalitis, and focal neurologic abnormalities can occur, including hemiparesis, cranial nerve palsies, and exaggerated deep tendon and/or pathologic reflexes. Patients may appear confused, agitated, or obtunded.

Results of imaging in patients with encephalitis may or may not demonstrate abnormal radiographic findings on CT or MRI modalities. CT scanning is useful to rule space-occupying lesions or brain abscess. MRI is sensitive for detecting demyelination, which may be seen in other clinical states presenting with mental status changes (eg. progressive multifocal leukoencephalopathy) and typical contrast enhancing (mostly temporal lesions).

Initial examination of the CSF, although not diagnostic, will usually confirm the presence of inflammatory disease of the CNS. The findings with encephalitis, aseptic meningitis and meningoencephalitis are generally indistinguishable.

5. SIH and fever

The fever seen in SIH cases those mimic meningoencephalitis might be explained by the release of pyrogenic cytokines by endothelial cells and astrocytes of blood-brain barrier secondary to a drop in CSF pressure [10]. These cytokines are the main mediators of inflammatory response in infectious and non-infectious disorders. Another suggested mechanism for fever would be an impaired hypothalamic thermoregulation secondary to mechanical distractions and venous engorgement in cavernous sinus and diencephalic region.

6. Neurologic manifestations of SIH

Hearing changes including disturbed sense of balance, tinnitus and echoing; visual changes including visual blurring and diplopia; various neurological symptoms including photophobia, phonophobia, amnesia and facial numbness are common [11]. A significantly decreased level of consciousness might be observed in cases with SIH and even they would admit with confusion and syncope [7]. Subtle cognitive deficits are common particularly during episodes of headache [2].

Symptoms in SIH patients related to the vestibulocochlear system such as disturbed sense of balance, tinnitus and sense of echoing may be explained by direct transmission of the abnormal CSF pressure to that in the perilymph [21]. Visual impairment was probably due to stretching of the cranial nerves due to downward displacement of the brain [12].

7. Radiological manifestations of SIH

CNS imaging is key in the differential diagnosis. Essentially no MRI findings are seen in aseptic meningitis and imaging is not needed in most of the cases. Cases with encephalitis have special MRI findings varying due to the etiology, being the temporal lobe involvement of HSV 1 encephalitis the most common. In contrary, MRI findings are characteristic and imaging with contrast material is preferred in SIH. With the wide-

spread and increasing use of MRI, SIH is more frequently recognized in recent years as an important cause of new onset persistent, daily, positional headaches. Diffuse pachymeningeal (dural) contrast enhancement is the main feature on cranial MRI. Some other findings of SIH have been described: Enlargement of pituitary, tenting of the optic chiasm, subdural fluid collections, engorged cerebral venous sinuses and findings due to sagging of the brain such as obliteration of subarachnoid cisterns, crowding of posterior fossa and descent of cerebellar tonsils [5].

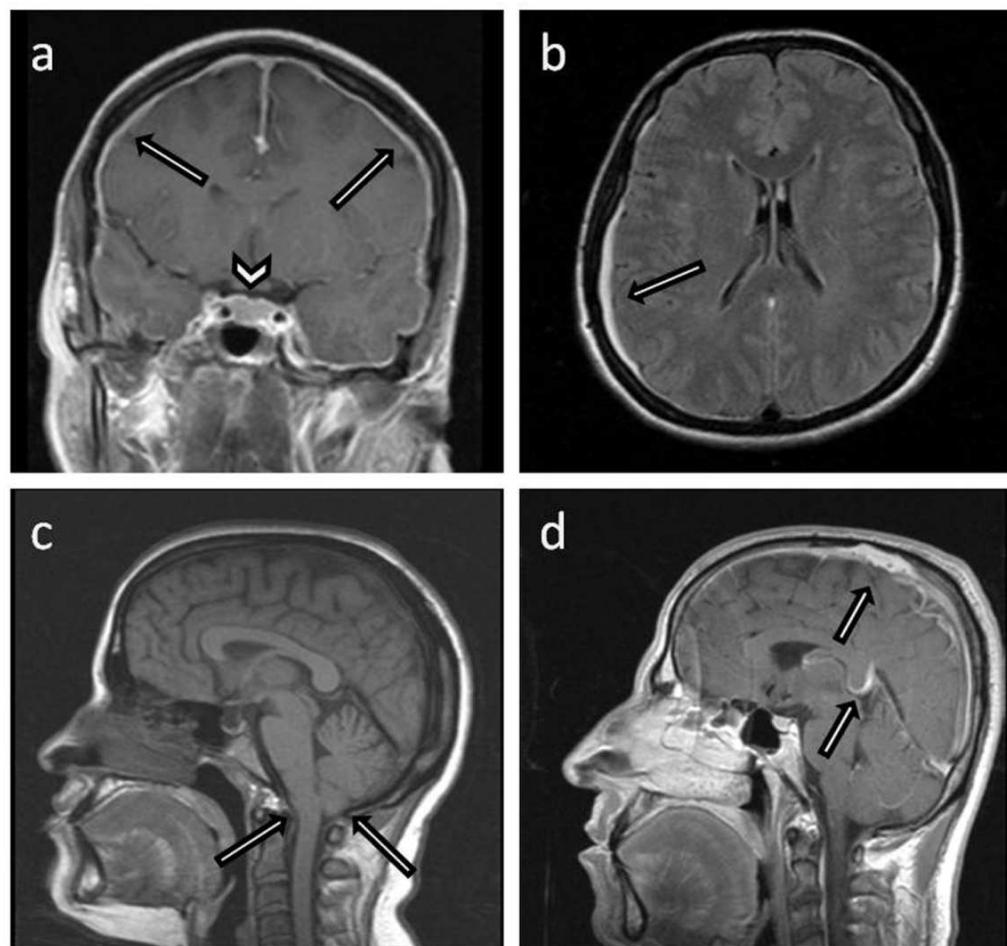


Figure 1. Thickening in meninges demonstrating pachymeningeal contrast enhancement (arrows) and enlargement of the pituitary gland (arrowhead) in a 36 years old female patient [7], presenting with orthostatic headache, stiff neck and fever. **b.** Axial FLAIR image of the same case showing bilateral thin subdural collection (arrow). **c.** Findings of sagging brain (arrow) in a 37 year old female (case 1] presenting with fever, severe headache, nausea and vomiting. **d.** Engorgement in cerebral veins (arrows) of a 44 year old male presenting with fever and meningeal irritation signs whose LP revealed a marked lymphocytic pleocytosis[450 / mm³]. (Derived from ref. 7. Photo courtesy of Sait Albayram, with permission)

8. Management and follow up

The treatment of SIH is controversial. In some, bed rest, hydration, caffeine intake, and steroids are effective, while in others an epidural blood patching may be needed [2,5]. Epidural blood patch is performed by obtaining a small amount of the patient's venous blood and injecting into the epidural space close to the site of the leakage; the resulting blood clot then patches the meningeal leak by forming a dural tamponade [9]. Although the outcomes have been poorly studied, epidural blood patch is the preferred modality for a better treatment outcome in those that do not improve with conventional supportive treatment [13].

9. Conclusion

Fever, headache, and meningeal irritation findings are generally accepted as the clinical features of meningitis syndrome. When CSF findings are not characteristically compatible with bacterial or tuberculosis meningitis, it is usually defined as aseptic meningitis. Some cases with SIH, those admitting with signs of meningeal irritation, decreased cognitive levels, and various neurological signs and symptoms including hyperexcitability and status, may mimic meningoencephalitis. The characteristic features of SIH should promptly be searched in those cases. If the headache is in orthostatic nature, CSF opening pressure is low and characteristic MRI findings are present, the diagnosis would be established as SIH. The diagnostic criteria defined by International Headache Society would be suggested to exclude SIH in the differential diagnosis.

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