Case Report

Thunderclap headache: It is always sub-arachnoid hemorrhage. Is it? – A case report and Review

Raghunath Aladakatti, Laxmikant B. Sannakki, Peter Y. Cai, Roselle Derequito

Department of Critical Care, Gopala Gowda Shanthaveri Memorial Hospitals, Mysore, Karnataka, India, Department of Neurology and Anesthesiology, University of Florida College of Medicine, Gainesville, Florida, USA

E-mail: *Raghunath Aladakatti - drraghunath.ha@gmail.com; Laxmikant Sannakki - bslaxmikant@gmail.com; Peter Y. Cai - petercai@ufl.edu; Roselle Derequito - rderequito@ufl.edu

*Corresponding author

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Abstract

Background: Spontaneous intracranial hypotension (SIH) is one of the relatively misdiagnosed pathophysiological entities by virtue of its presentation. SIH is a condition involving reduced intracranial pressure usually secondary to dural tear. There is recent increase in reporting of its varied presentation in literature. Critical care physicians and neurosurgeons are recognizing it in higher numbers than before. SIH is characterized by sudden onset of orthostatic headache and may be associated with neck stiffness, nausea, vomiting, tinnitus, deafness, and cognitive abnormalities. Since its imaging characteristics resemble classic subdural hematoma from other causes wrong diagnosis and intervention might have devastating outcome.

Case Description: Here we discuss a case presented to us with severe headache of sudden onset without any associated problems. Patient was initially being treated as sinusitis and later diagnosed as bilateral subdural hematoma and surgical intervention was being considered. Thorough history taking and physical examination lead to strong suspicion of intracranial hypotension (IH) and patient showed dramatic improvement with epidural blood patch.

Conclusion: SIH is a commonly misdiagnosed entity. A high index of suspicion is required for timely diagnosis, in order to minimize unwanted therapeutic interventions that can worsen the patient’s condition and to help initiate early and simple interventions.

Key Words: Diagnosis, hypotension, intracranial, presentation, spontaneous

INTRODUCTION

The earliest reported case of symptoms as a result of intracranial hypotension (IH) was back in 1825 when vertigo and unsteadiness was described in a patient after removal of cerebrospinal fluid (CSF). Later in the 20th century, patients with headaches secondary to lumbar puncture (LP) were identified due to this entity.[12,14] IH, initially described in 1938, can mimic many neurological syndromes by virtue of its presentation and as a result, is frequently misdiagnosed.[15] Spontaneous intracranial hypotension (SIH) was first described as a potential cause of postural headache, and the term “spontaneous aliquorrhea” was coined by Schaltenbrand.[17]
Subsequently, IH was classified into many categories as per the underlying etiologies: Primary or spontaneous, post-LP, following head injury or craniotomy, and hypovolemia induced.[1] The estimated prevalence of IH is 1 in 50,000 adults.[2] Common clinical features are orthostatic headache, seizure, altered mental status, neck stiffness, nausea, double vision, dizziness, hearing impairment, visual blurring, photophobia, visual field defect, facial weakness, ataxia, bulbar weakness, stupor, and other encephalopathy features.[3] Decreased CSF pressure results in decreased buoyancy and “brain sagging” leading to traction on the leptomeninges and neural structures.[4] This traction is believed to be the main cause of headache and other neurological symptoms found in SIH. Dural venous engorgement may also be another source of symptoms.[5] Patients with SIH are often treated for their subdural fluid collections or tonsillar herniation instead of correcting the underlying cause, IH. This report presents a patient, who was successfully treated for SIH, in the context of clinical presentation, diagnosis, and treatment approach.

**CASE HISTORY**

A 38-year-old Asian male presented with “sudden onset of worst headache of my life” that was generalized in nature and not associated with nausea, vomiting, or seizures. His past history was unremarkable. He was initially treated for sinusitis and prescribed a course of antibiotics and nasal decongestants, which initially improved his symptoms. After approximately one month, the patient’s headache reoccurred and persisted. He was evaluated in the emergency room for recurrent headache and urgent computed tomography (CT) brain revealed bilateral subdural collections (images not shown). With the diagnosis of bilateral subdural hematoma, he was considered for possible surgical intervention. Later careful history analysis revealed that his headache was worst when standing or sitting, but relieved when lying down. Other causes of headache were ruled out. On examination, all vital signs were within normal limits. There was no neck stiffness and no rash. Systemic examination was unremarkable. On neurological evaluation, patient was alert, oriented with intact higher cognitive function including speech. Cranial nerves including fundus exam, motor, sensory, and cerebellar examination demonstrated nonfocal neurological exam and no abnormality detected. Considering the patient’s age, symptoms, unremarkable past medical problems, nonfocal exam, and radiological findings prompted us to consider SIH as the diagnosis and magnetic resonance imaging (MRI) brain was ordered to confirm it. MRI brain revealed [Figures 1 and 2] the classic features of IH including subdural hematoma collection, descent of cerebellar tonsils, effacement of the prepontine cistern and interpeduncular cistern, inferior displacement of the optic chiasm, reduced mammillary body and pontine distance due to the descent of the mammillary body, crowding of posterior fossa due to brainstem descent, descent of cerebellar tonsils, sagging of tuber cinereum, inferior displacement of splenium, and descent of the fastigium of the fourth ventricle.

MRI myelography showed epidural fluid at the C2-C4 level with possible dural tear at C2 on right side and C3, C4 on the right side. Epidural fluid was also seen in the entire dorsal region and in the upper lumbar region without any obvious dural tear. The patient was scheduled for an epidural patch, which was performed at the cervical level using 12 cc of autologous blood. Symptoms resolved significantly after the epidural blood patch. Repeat CT head 3 months postblood patch showed resolution of the subdural fluid collection [Figure 3]. Patient is asymptomatic till date.
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DISCUSSION

In 1942, the first case of IH was reported by Puech.[11] Trauma, LP, or surgery has been recognized as possible causes of IH since the early 1900s.[1] Headache, vertigo, nausea, convulsions, stupor, and death from “cerebral hypotension” were documented by Leriche and Wertheimer in patients following LPs, head trauma, cranial operations, or unrelated medical or neurological disorders.[6,12] SIH, however, has been poorly characterized and diagnosed. The predominant symptom of this syndrome is postural headache of acute onset, which is mild when the patient is lying down but rapidly worsens when the patient sits or stands. The pain experience is described as a ‘thunderclap headache,’ similar to subarachnoid hemorrhage patients. Other symptoms include neck pain and stiffness, diplopia, nausea, vomiting, vertigo, tinnitus, impaired hearing, convulsions, and cognitive abnormalities.[5] Coma and encephalopathy have also been noted.[25] IH can also mimic nonconvulsive status epilepticus clinically and electrographically.[4]

Diagnosis

The International Classification of Headache Disorders has developed diagnostic criteria for SIH [Table 1].[27] Most cases of SIH are not due to dramatic rupture of arachnoid cysts but rather due to spontaneous slow leakage of CSF via small dural defects. The precise mechanism by which these dural defects arise is unknown but they are thought to represent an underlying weakness in the structural integrity of the meninges.[23] Connective tissue disorders account for up to two-thirds of patients with intracerebral hemorrhage (ICH).[21] The diagnosis is most often confirmed by cranial MRI scanning, where the characteristic feature is subdural fluid collections (50% of cases).[22] Characteristic brain MRI changes can be best remembered by the mnemonic “SEEPS” for Subdural fluid collections, pachymeningeal Enhancement, Engorgement of venous structures, Pituitary hyperemia and Sagging of the brain.[20] The earliest finding is distension of the dural sinuses. Brain stem herniation is seen in late or severe cases. Other features include diffuse continuous linear pachymeningeal enhancement without nodularity both above and below the tentorium, reduced distance between the pons and the mammillary body due to the descent of the mammillary body, crowding of posterior fossa due to brainstem descent, bilateral or unilateral subdural fluid collection, pituitary enlargement, descent of cerebellar tonsils, effacement of the preoptic cistern and interpeduncular cistern, inferior displacement of the optic chiasm, sagging of tuber cinereum, inferior displacement of splenium, slit-like ventricles, descent of fastigium of the fourth ventricle and decreased ventricular size.[4] Epidemiological data suggests that the incidence of SIH is approaching that of subarachnoid hemorrhage.[25] Other modalities such as CT myelography, spinal MRI, radio nucleotide cisternography, and Doppler flow imaging also are useful diagnostic tools.[2,8,10,28]

![Figure 3: Postprocedure CT head: Noncontrast CT head 3 months after the blood patch shows complete resolution of the subdural hematoma](image)

**Table 1: International classification of headache disorders SIH criteria**

| At least one of the following and fulfilling criterion D | At least one of the following | Must be fulfilled | Must be fulfilled |
|--------------------------------------------------------|-------------------------------|------------------|------------------|
| Neck stiffness                                         | Evidence of low CSF pressure on MRI (e.g., pachymeningeal enhancement) | No history of dural puncture | Headache resolves within 72 h or other cause of CSF fistula after epidural blood patching |
| Tinnitus                                               | Evidence of CSF leakage on conventional myelography, CT myelography or cisternography |                      |                  |
| Hypacusia                                              | CSF opening pressure <60 mm H₂O in sitting position |                  |                  |
| Photophobia                                            |                               |                  |                  |
| Nausea                                                 |                               |                  |                  |

Treatment

The mainstay of SIH treatment is restoring the CSF pressure and relieving symptoms. There are multiple treatment theories that aim to achieve these goals, such as conservative medical therapy, epidural or intrathecal injections and surgery. Medical therapy, which includes bed rest, oral hydration, caffeine, and steroids, can be planned for patients with mild symptoms.[13,15] Despite many treatment modalities available, there are no randomized control trials assessing their efficacies and risks. As a result, conservative treatment is the first measure in mild symptoms. If the symptoms persist or
some experts use multiple. A total of 10-35 ml of autologous blood is injected into the epidural space and patients are placed in trendelenburg position for an hour. If this is unsuccessful, up to 100 ml of autologous blood can be used. Approximately 30-70% of patients improve after the first blood patch and 30-50% of the remainder improves after the second patch,[5,26] some experts use multiple injection sites if the leakage is at different levels or in resistant or recurrent cases. Epidural injection of fibrin glue has also shown promising results for treatment of SIH. However, this therapy requires exact localization of the leak in order to be successful. In patients who do not respond to treatment and have definite site of CSF leak, a surgical repair can be pursued. This includes meningeal diverticula repair, which involves packing CSF leaks with muscle or gel foam soaked with blood.[7,19] Interestingly, in some cases, imaging abnormalities remained despite clinical recovery.[5]

**CONCLUSION**

SIH is a commonly misdiagnosed entity. A high index of suspicion is required for timely diagnosis, in order to minimize unwanted therapeutic interventions that can worsen the patient’s condition and to help initiate early and simple interventions.

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