Spontaneous Intracranial Hypotension Presenting Without Orthostatic Headache Complicated by Acute Subdural Hematoma After Drainage for Chronic Subdural Hematoma
—Case Report—

Takumi KURAMAE,1 Joji INAMASU,1 Yu NAKAGAWA,1 and Masashi NAKATSUKASA1

1Department of Neurosurgery, Saiseikai Utsunomiya Hospital, Utsunomiya, Tochigi

Abstract
A 28-year-old man presented with a case of spontaneous intracranial hypotension (SIH) manifesting as a bilateral chronic subdural hematoma (CSDH) without orthostatic headache. He developed life-threatening acute SDH as a complication of CSDH drainage. Neurosurgeons should be aware that SIH patients do not always present with orthostatic headache. Brain magnetic resonance imaging with gadolinium may be recommended for young adults with non-traumatic CSDH before drainage to exclude SIH, even if they do not present with orthostatic headache.

Key words: chronic subdural hematoma, complication, drainage, spontaneous intracranial hypotension, orthostatic headache

Introduction
Diagnosis of spontaneous intracranial hypotension (SIH) may be established without much difficulty in patients with chronic headache, particularly if the medical history includes complaints of orthostatic headache.5,7,16 SIH is often associated with, and may be the cause of, non-traumatic chronic subdural hematoma (CSDH).5,7,16 So CSDH patients with underlying SIH must be differentiated from those without SIH: if presence of SIH remains unnoticed, CSDH may recur despite repeated drainages.2,4,11,19 Few serious complications have been reported, however, after drainage for CSDH associated with SIH.2,4,11,19 We present a rare case of SIH with concomitant CSDH in which life-
threatening acute SDH developed after CSDH drainage.

Case Report

A previously healthy 28-year-old man was brought to our clinic by his parents. He was mildly disoriented with a consciousness level of G4V4M6 on the Glasgow Coma Scale (GCS). No other focal neurological deficits were found. He complained of worsening frontal headache which did not change with posture. He denied previous trauma to the head and neck, and he could not recall when the headache had started. Brain computed tomography (CT) revealed a bilateral CSDH compressing the adjacent brain (Fig. 1), and he was brought to the operating room for drainage of the CSDH later on the day. Although the preoperative differential diagnosis included SIH, this was considered unlikely because of the absence of orthostatic headache.

He underwent bilateral burr-hole drainage of the CSDH. Although the procedure was accomplished uneventfully, he became comatose approximately 12 hours later, with dilated right pupil. CT showed an acute SDH on the right with marked brain shift (Fig. 2), which was considered responsible for the deterioration. The acute SDH was removed through an emergency craniotomy, but he remained stuporous with a GCS score of E2V3M5.

Suspicion for SIH as an underlying cause of CSDH grew stronger, and magnetic resonance (MR) imaging of the brain with gadolinium was obtained 1 day after the craniotomy, which revealed diffuse meningeal enhancement, establishing the diagnosis of SIH (Fig. 3). Subsequently, CT myelography was performed, and revealed cerebrospinal fluid (CSF) leakage at the C1-2 level (Fig. 4). Epidural blood patch (EBP) was performed to terminate the leakage through a Tuohy needle placed from the C6-7 interlaminar space, but his neurological condition failed to improve. We thought that the possible dural tear might be too large for EBP to terminate the CSF leakage, and 2
days after EBP, spine surgery to seal the leakage at C1-2 was performed.

After C1 laminectomy, the CSF leakage though the lacerated dura at C1-2 was visible without Valsalva maneuver. No meningeal cysts or other anomalies around the spinal nerve roots were present under the operating microscope. Subsequently, the C1-2 epidural space was packed tightly with pieces of muscle and fibrin glue. Postoperative course was uneventful, and he was alert and oriented on the day after spine surgery. Follow-up CT obtained 10 days after spine surgery showed resolution of the CSDH (Fig. 5), and he was discharged 2 weeks after spine surgery without neurological deficits. He remained neurologically intact after 12 months.

Discussion
Orthostatic headache is the hallmark symptom of SIH, and is so characteristic of the disorder that its presence is considered prerequisite for the diagnosis, as defined by the International Headache Society.) However, neurosurgeons should be aware that not all patients with orthostatic headache have underlying SIH, and not all patients with SIH present with orthostatic headache. The sensitivity/specificity of orthostatic headache as a diagnostic indicator for SIH remains unknown. Interestingly, orthostatic headache is not prerequisite in the new proposed diagnostic criteria for SIH. SIH patients presenting with altered mental status or dementia often reported constant rather than orthostatic headache. Such altered mental status may prevent the patients from describing the characteristics of their headache correctly. Another possibility is that SIH patients who present with altered mental status may have a larger dural hole compared with those who are neurologically intact. Consequently, CSF leakage in the former patients may not subside even in the recumbent position, and as a consequence, the headache persists regardless of the posture. Moreover, larger amount of CSF leakage may result in rapid downward displacement of the brain, brainstem compression, and altered metal status. In the present case, continuous CSF leakage from the lacerated dura at C1-2 was visible intraoperatively. The failure of the EBP may be explained by the large dural laceration at C1-2. Whether medically intractable CSF leakage at C1-2 is common remains controversial, but spine surgery or other special techniques for EBP to stop the leakage are efficacious in cases refractory to ordinary EBP from the lumbar spine.

SIH patients with concomitant CSDH seem to complain of orthostatic headache less frequently than those without CSDH. Therefore, the presence of CSDH may prevent orthostatic headache from developing by correcting the abnormally low intracranial pressure (ICP) or volume. In the present case, the headache was not orthostatic, so we assumed incorrectly that SDH was unrelated to SIH and performed drainage which resulted in the complication of acute SDH. If brain MR imaging with gadolinium had been obtained before CSDH drainage, we might have established the correct diagnosis of SIH. Our experience highlights the need to suspect SIH in young adults with non-traumatic CSDH, even if the headache is not orthostatic.

Delay in the diagnosis of SIH is not uncommon, and patients with CSDH may be the most vulnerable: SIH is often associated with CSDH on imaging studies, and SIH patients may have undergone repeated drainages before definitive diagnosis is made. The incidence of CSDH among SIH patients ranges from 20% to 40%. Most misdiagnosed SIH patients with CSDH remain neurologically intact despite repeated drainages, and severe complication is rare, such as the postoperative acute SDH in our case. In retrospect, bilateral drainage of the CSDH in the present case may have led to sharp fall in the ICP, traction and disruption of bridging veins, and finally acute bleeding. However, the bleeding source was not identified during craniotomy for acute SDH evacuation, so this explanation remains speculative.

No SIH management guidelines are accepted worldwide, and in particular, the sequence of treatment remains undefined in patients with concomitant CSDH. Measures to stop CSF leakage, particularly EBP, are required to precede CSDH drainage. Small CSDH may resolve spontaneously after successful treatment of SIH. Nevertheless, CSDH drainage may have to be performed emergently in patients with impending brain herniation. To complicate matters, cases of SIH that resolved spontaneously after CSDH drainage have been reported. More efforts to establish the sequence of treatment for SIH patients with concomitant CSDH is needed.

References

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Diagnostic Pitfall in SIH Without Orthostatic Headache


Address reprint requests to: Joji Inamasu, MD, Department of Neurosurgery, Saiseikai Utsunomiya Hospital, 911-1 Takebayashi, Utsunomiya, Tochigi 321-0074, Japan. e-mail: ginamasu@aol.com