Spontaneous intracranial hypotension (SIH) syndrome, formerly known as aliquorrhea, is characterized by orthostatic headache that improves rapidly when the patient is recumbent. According to the Monro-Kellie hypothesis, SIH is usually a consequence of spontaneous cerebrospinal fluid (CSF) leakage that results in a volume imbalance of CSF and intracranial blood in the brain [1]. Most patients have a benign clinical course; however, some patients suffer from chronic or acute subdural hematoma (SDH) owing to downward displacement of the brain [2–4]. Sagging of the brain can affect consciousness owing to acute diencephalic compression [5]. We present our experience of treating two patients in whom SIH caused complications but who recovered successfully after burr hole drainage.

Key Words: intracranial hypotension syndrome, intracranial pressure, subdural hematoma, trephination

Spontaneous intracranial hypotension is a potentially severe condition that is caused by continuous cerebrospinal fluid leakage. Clinically, most patients have a benign course and the condition remits after conservative management. We report two consecutive patients who presented with acute expansion of subdural collection and disturbed consciousness. Both patients recovered completely after undergoing burr hole drainage.

CASE PRESENTATIONS

Case 1
A 35-year-old man suffered from intermittent clear rhinorrhea for 2 weeks. He developed a severe orthostatic headache 2 weeks after the nasal discharge ceased. He denied any history of trauma although he had been mountain climbing 1 month before admission. Lumbar puncture revealed normal CSF opening pressure and normal biochemistry. Computed tomography (CT) scanning and magnetic resonance imaging (MRI) of the brain revealed bilateral subdural fluid collection and a sagging brain typical of SIH (Figure 1). Two days after admission, the patient became stuporous. CT revealed a mildly enlarged left SDH, and emergency trephination was performed (Figure 2A). Bloody subdural fluid with initial high pressure and a sagging cerebral cortex were noted during the operation.

The patient regained consciousness after trephination; however, he became stuporous again 2 days later. Repeated brain CT showed that the right SDH had become larger. Therefore, a second trephination was performed on the right side. Similar operative
findings were noted and an external ventricular drain (EVD) was inserted into the subdural cavity for both continuous drainage and intracranial pressure (ICP) monitoring. The patient regained consciousness immediately, and the ICP was negative over the following 3 days. Examination of the subdural fluid revealed a high red blood cell (RBC) content (RBC count, $1.56 \times 10^6/\mu L$; white blood cell count, $1,780/\mu L$), which indicated acute bleeding in the subdural space.

Three weeks after admission, the patient became drowsy and developed urinary incontinence. Brain CT revealed left chronic SDH with brain shift (Figure 2B). Therefore, the patient underwent trephination for the third time. He recovered completely, and no neurologic deficits were noted at follow-up 2 years later. Magnetic resonance myelography and radioisotope cisternography did not demonstrate any evidence of CSF leakage. Follow-up MRI 1 year after the operation revealed that the pachymeningeal enhancement, subdural fluid collection and downward brain shift had disappeared.

**Case 2**

A 31-year-old man had a 3-month history of progressive positional headache associated with symptoms of nausea, vomiting and mild blurred vision. The diagnosis of SIH was confirmed by lumbar puncture (opening pressure, $5 \text{cmH}_2\text{O}$; normal cell count and
biochemistry) and MRI findings (diffuse pachymeningeal enhancement; Figure 3A). Conservative treatment with hydration was initially given, but the patient’s symptoms persisted. Three months later, he developed acute onset of slurred speech, explosive orthostatic headache, dysarthria and posterior neck pain. Follow-up brain MRI revealed a chronic left SDH with brain shift (Figure 3B). The patient underwent burr hole drainage. Initial high pressure with xanthochromic subdural fluid was noted while performing trephination. The patient recovered well and his headache improved progressively.

DISCUSSION

Intracranial hypotension syndrome is generally a benign process, and most cases are responsive to conservative treatment. The headache typically resolves with bed rest and hydration. Lumbar epidural blood patches or epidural infusion of saline may provide alternative and effective treatment modalities [6,7]. With the improvement of imaging modalities, atypical disabling presentations are increasingly being recognized, including parkinsonism, frontotemporal dementia, syringomyelia, hypopituitarism, seizures, coma, and even death [8].

Despite the term “spontaneous” intracranial hypotension, many authors think that mechanical stress plays an important role in the pathogenesis of SIH by acting on the focal weak point in the dural sac, the most common leak site identified in SIH patients [9]. Many patients with SIH present without known predisposing factors or a history of major trauma; however, it has been reported that some patients present with SIH after having sustained trivial trauma [10], or accompanied by meningeal diverticula [11], or connective tissue disorders [12,13]. Chung et al reported that during a detailed interview, seven (23%) of 30 patients recalled a prior trivial neck or head trauma [10]. However, our patients could not retrospectively recall any trauma history, and imaging findings failed to demonstrate any evidence of CSF leakage. Although nasal CSF rhinorrhea after mountain climbing was suspected in the first patient, his symptom of positional headache began 2 weeks after cessation of the nasal discharge. Notably, no evidence of traumatic CSF leakage or dural defect could be identified from serial imaging studies.

The diagnosis of SIH depends on the clinical history, imaging findings, and characteristics of CSF studies. Low opening pressure on lumbar puncture (usually <6 cmH₂O) is a hallmark characteristic in patients suspected of having SIH [9]. The reported incidence of subdural collections in association with SIH is approximately 10% [14], and lumbar puncture may be contraindicated in these patients because of a possible life-threatening downward brain shift [15]. In Case 1, the lumbar puncture was performed carefully for diagnosis because of an equivocal clinical history and imaging findings. Nevertheless, the patient developed acute deterioration of consciousness owing to
enlargement of the subdural collection 2 days after the procedure. We suggest that lumbar puncture may be contraindicated in SIH patients with subdural collections and brain shift, and this procedure should be performed with caution.

Chronic SDH is an uncommon sequela of SIH [3,4], and patients with this sequela, including those described here, usually have a good clinical outcome after simple burr hole drainage. de Noronha et al reported on four consecutive SIH patients with acute deterioration of consciousness owing to enlarged subdural collections; all of them had favorable outcomes after surgical drainage [14]. It is difficult, however, to distinguish whether these patients have high or low ICP because both conditions exhibit similar imaging findings (downward displacement of brain with subdural fluid collections) and clinical symptoms such as headache and consciousness disturbance. Sayer et al reported that one patient with SIH became comatose after repeated burr hole drainage of bilateral subdural fluid collection [16]. The patient had a subdural hygroma without obvious signs of increased intracranial hypertension. Therefore, they concluded that burr hole drainage in such a complicated SIH patient in whom there was no evidence of elevated ICP may be harmful and that trephination should be performed with caution.

In our first patient, however, the operative findings supported our initial suspicion that the initial increased ICP was related to acute expansion of the subdural fluid volume. The high RBC content in the subdural fluid suggested acute bleeding within the subdural space. The fact that the patient regained consciousness immediately after burr hole drainage also supported the preoperative increased ICP and justified the timing for surgical evacuation of the SDH. Subdural placement of an EVD tube also provided a useful treatment option for ICP monitoring and delineation of subdural fluid content. In addition, this provided another way to confirm the diagnosis of SIH other than by clinical and imaging findings.

Although SIH is usually a benign process, a complicated condition such as chronic or acute SDH with acutely increased ICP should be promptly recognized, especially when the patient’s consciousness has deteriorated. Subdural placement of an EVD both for ICP monitoring and drainage may be helpful in such complicated cases.

REFERENCES

以顱骨鑽孔引流手術治療複雜性之顱內低壓症候群
— 兩病例報告

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自發性顱內低壓症候群起因於連續性之腦脊髓液流失，是一具有潛在危險性之病症。
臨床上，大部分患者經保守治療後均可痊癒。本文報告二位病例因合併意識惡化以及
急性硬腦膜積液之擴大，在接受顱骨鑽孔引流手術後，均獲完全的康復。

關鍵詞：顱內低壓症候群、顱內壓、硬腦膜下血腫、顱骨鑽孔術

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