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Clinical Article

Spontaneous Intracranial Hypotension : Clinical Presentation, Imaging Features and Treatment

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Objective : In the present study, the authors investigated the clinical and imaging features as well as the therapeutic outcomes of SIH (spontaneous intracranial hypotension) patients.
Methods : A retrospective review of 12 SIH patients was carried out. The diagnostic work-up included lumbar tapping and measurement of CSF opening pressure, radioisotope cisternography, brain and spinal magnetic resonance imaging (MRI), and computed tomography (CT) myelography. Autologous epidural blood patching was performed in patients who did not respond to conservative therapies, including analgesics, steroids, hydration and rest.
Results : Typical postural headache was found in 11 (91%) patients. Nine (75%) patients showed pachymeningeal enhancement on their initial T1-weighted MR images. The CSF opening pressure was less than 60 mmH_2O in 9 of 11 patients. Autologous epidural blood patching was performed in 7 patients, and all of them showed good responses.
Conclusion : SIH can present with various clinical presentations and neuroimaging findings. Autologous epidural blood patching is thought to be the treatment of choice for patients with SIH.

KEY WORDS : Headache · Cerebrospinal fluid leak · Dural enhancement · Magnetic resonance imaging · Blood patch · Intracranial hypotension.

INTRODUCTION

Although spontaneous intracranial hypotension (SIH) is rare, it has become increasingly recognized in recent years, and it is now a well-known disorder. SIH is characterized by the diagnostic triad of postural headache, diffuse pachymeningeal gadolinium enhancement, and low cerebrospinal fluid (CSF) opening pressure, below 60 mmH_2O[13]. However, several types of SIH patients have recently been reported, including patients with normal CSF pressure, absence of pachymeningeal enhancement or non-orthostatic headache[6,8,10,12]. Therefore, the authors investigated the clinical and imaging features as well as the therapeutic outcomes of patients with SIH.

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MATERIALS AND METHODS

We retrospectively reviewed the medical records and telephone interview notes of 12 patients with SIH who visited our institution between June 1995 and February 2008. We excluded patients with history of craniospinal surgery, epidural anesthesia, spinal tapping, and spinal trauma. The patients ranged in age from 29 to 53 years. There were 9 female and 3 male patients. They visited our hospital due to characteristic headaches that worsened in the upright position. All of the patients underwent brain magnetic resonance imaging (MRI) with gadolinium enhancement. Under the suspected diagnosis of SIH, radioisotope cisternography was performed in 11 of the 12 patients and computed tomography (CT)-myelography was performed in one patient in an effort to find evidence of CSF leakage. CSF tapping was performed to measure the opening pressure and analyze the elements. Spine MRI was performed in 3 patients.

Autologous epidural blood patching was performed in patients who did not respond to conservative therapy. For

performing epidural blood patching, an 18-gauge needle was inserted into the epidural space by the loss of resistance technique under prone position at the site where the CSF leakage was suspected. The needle was identitfied in the epidural space by contrast material injection under X-ray monitoring[18]. Ten to twenty milliliters of autologous venous blood was slowly injected via this needle.

RESULTS

The clinical and radiological findings of the patients with SIH are summarized in Table 1, along with the treatment modalities used. All of the patients had typical orthostatic headaches, except for one demonstrating chronic lingering type of headache with intermittent nausea, vomiting regardless of positional change. Additional clinical manifestations were nausea, nuchal pain, vomiting, pain in the interscapular area, dizziness, tinnitus, nystagmus, ear fullness, and blurred vision. The mean symptom duration prior to admission was 19 days (range of 2-39).

Table 1. Demographic, clinical, imaging characteristics, and treatment of 12 pateints with SIH

Variable	No. of Patients
Age (years), range (mean±SD)	29-53 (40±6.9)
Male/female	3/9
Precipitating event (n=8)	
Lifting a heavy thing	3 (25%)
URI with cough	2 (16%)
Dental care	1 (8%)
Singing with handclap	1 (8%)
Rope jumping	1 (8%)
Symptom (n=12)	
Postural headache	11 (92%)
Nausea	9 (75%)
Vomitting	4 (33%)
Nuchal pain	5 (33%)
Interscapular pain	3 (25%)
Blurring of vision	1 (8%)
CSF (n=11)	
Lumbar opening pressure (mmH_2O) : range (mean±SD)	30-80 (52.2±15.6)
<60 mmH_2O	9 (81%)
>60 mmH_2O	2 (19%)
WBC (no./mm^3) : range (mean±SD)	0-10 (3.7±3.1)
RBC (no./mm^3) : range (mean±SD)	0-1125 (281.9±381.2)
Glucose (mg/dL) : range (mean±SD)	48-91 (63.2±12.8)
Pretein (mg/dL) : range (mean±SD)	43-110.9 (66.95±24.2)
Brain MRI (n=12)	
Diffuse pachymeningeal enhancement	10 (83%)
Early stage subacute SDH	1 (8%)
Subdural hygroma	1 (8%)
Radioisotope cisternography (n=11)	
Leakage	6 (54%)
Delayed migration of radioactivity over cerebral convexities	7 (63%)
Treatment (n=12)	
Conservative manage	5 (42%)
Epidural patch	7 (58%)

SIH : spontaneous intracranial hypotension, SD : standard deviation, MRI : magnetic resonance imaging, WBC : white blood cell, RBC : red blood cell, SDH : subdural hematoma, CSF : cerebrospinal fluid

CSF analysis revealed no pleocytosis (more than 10 cells/mm^3). The mean WBC cell count was 3.7 cells/mm^3. The mean CSF glucose concentration was 63.2 (mg/dL) (range of 48-91). Protein was within normal limits with a mean value of 66.9 (mg/dL). CSF opening pressure could not be measured in two patients, even after several trials. Two patients had an opening pressure over 60 mmH_2O . The mean value of opening pressure was 52.2 mmH_2O (range of 30-80 mmH_2O). An exceptional case without postural headache also showed low CSF opening pressure.

The major MRI feature of diffuse pachymeningeal gadolinium enhancement was observed in 9 of 12 patients. One of the three patients who did not demonstrate pachymeningeal enhancement on the initial study showed delayed dural enhancement four days after epidural blood patching. Bilateral subdural fluid collection was noted in one patient. Radioisotope cisternography demonstrated leakage sites in five cases. Delayed migration of radioactivity over the cerebral convexities, even at 24 hours post-injection, was noted in seven cases. Early appearance of radioactivity in the kidneys and urinary bladder was observed in three cases. CT myelography was performed in one patient, and it demonstrated the actual site of CSF leakage. Four patients underwent spine MRI. Epidural free fluid from thoracic to lumbar area and abnormal epidural venous engorgement in the lumbar area were demonstrated in one patient.

All cases were initially treated with conservative therapies, including bed rest, intravenous fluid infusion, and oral analgesics. Oral glucocorticoid therapy and caffeine treatment were attempted in two patients, but the effects were questionable. Autologous epidural blood patching was performed in seven patients who did not respond to conservative therapy. All of the patients responded well to epidural blood patching.

Illustrative case

A 35-year-old woman presented with a 2-week history of headache, nausea, vomiting, and nuchal pain. The headache was so severe in the standing position that she could not stand up and walk for just 10 seconds. The findings of all general medical and neurological examinations were normal. MRI studies of the head showed diffuse pachymeningeal gadolinium enhancement. Lumbar punctures were performed with immeasurable opening pressure. On

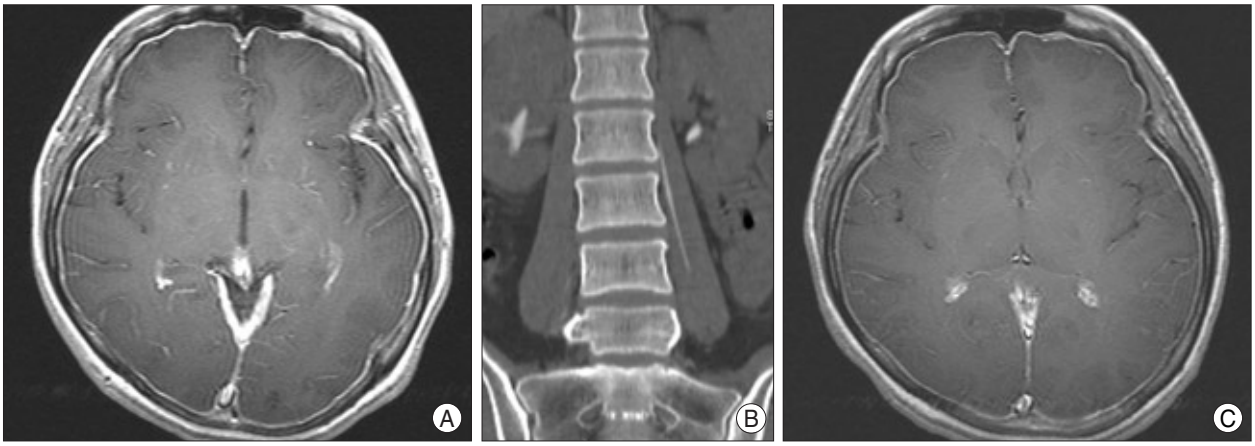

Fig. 1. A : Axial gadolinium-enhanced T1-weighted magnetic resonance image shows intense diffuse pachymeningeal enhancement. B : Computed tomography-myelography showing abnormal contrast extravasation in the psoas muscle. C : Follow-up image shows some resolution of meningeal enhancement on the third day after epidural patching.

CSF analysis, WBC cell count was 6 cells/mm^3, and the protein count was 96.5 mg/dL. CT myelography of the lumbar spine showed abnormal contrast extravasation at the level of L4, 5, S1 from the nerve root to the psoas muscle (Fig. 1).

Although the patient was initially treated with conservative therapy consisting of bed rest, intravenous fluid infusion, and oral analgesics, she did not respond sufficiently to the conservative therapy. The patient recovered well after the administration of 12 cc of autologous epidural blood patch, and she was able to lead a normal life. Brain MRI revealed some resolution of meningeal enhancement 3 days after epidural blood patching.

DISCUSSION

SIHs are rarely encountered in neurological practice. However, there have been numerous reports of SIH over the last decade, and it is now a well-known disease entity. The classical diagnostic triad of SIH includes postural headache, diffuse pachymeningeal gadolinium enhancement, and low CSF opening pressure, below 60 mmH_2O[9]. There have been many recent reports concerning several variant types of clinical manifestations of SIH, including (1) normal range of CSF opening pressures with typical postural headache and diffuse pachymeningeal enhancement on MRI, (2) absence of diffuse pachymeningeal gadolinium enhancement with the presence of low CSF pressure and typical clinical manifestations, and (3) absence of headaches despite low CSF pressure and the presence of diffuse pachymeningeal gadolinium enhancement[8,10-12].

SIH patients exhibit a wide variety of symptoms in addition to headache with postural change, including nausea, vomiting, nuchal pain, pain in the interscapular area, blurred vision, tinnitus, dizziness, nystagmus, ear fullness, photophobia, and phonophobia. It was reported that the headache could be non-postural[5,6], or even absent[11] in SIH patients. In our series, we also had a patient whose headache was the chronic lingering type. However, the possibility of SIH could not be completely ruled solely on the basis of the non-postural nature of the headache. In the previous reports, patients with non-postural headaches showed gradual improvement with conservative therapy[6].

Since diffuse pachymeningeal gadolinium enhancement was first reported to be the most characteristic neuroimaging feature of intracranial hypotension in 1991[8], many investigators have reported additional cranial MRI features of SIH, including brain sagging, pseudo-Chiari malformation (descent of the cerebellar tonsils sometimes mimicking Chiari I malformation[1,14,15]), and subdural fluid collection[2,7]. Caudal displacement of the cerebellar tonsils is generally mild and rarely extends more than several millimeters below the level of the foramen magnum[16]. There have been recent reports of intracranial hypotension without diffuse pachymeningeal enhancement[4,10,17]. The possibility that the timing of the MRI determines the presence or absence of diffuse pachymeningeal enhancement was reported by Fuh et al. in 2007[4]. In our series, three of nine (33%) patients did not demonstrate pachymeningeal enhancement on the initial study but had postural headaches with low lumbar opening pressure. Therefore, we recommend that invasive studies, such as lumbar puncture including measurement of lumbar opening pressure, should be done in patients with clinically typical postural headache but normal findings on MRI for the diagnosis of SIH.

It has been well-known that the fundamental pathophysiologic conception of SIH is CSF leakage resulting in CSF hypovolemia with low CSF opening pressure, however, two

of the patients (16.7%) in our series had an opening pressure over 60 mmH_2O. This exceptional phenomenon was reported as another type of SIH or result of a compensatory response to CSF leakage[6]. Another hypothesis is that certain volume loss may occur without a low pressure or that the check-valve phenomenon might occur when a pressure- or volume-dependent CSF leakage allows CSF leak only at higher volume or pressure[12]. Although the etiology of spontaneous CSF leaks remains elusive in the majority of patients, the possibility of a pre-existing weakness of the dural sac, such as an arachnoid diverticula or dural diverticula, has been suggested as a plausible hypothesis[3]. Spinal meningeal diverticula are abnormal outpouchings of the common dural sac, the spinal arachnoid, or the nerve root sheath. In some cases, several heritable connective tissue disorders, including Marfan's syndrome and neurofibromatosis 1, have been associated with spinal diverticula[16]. It is important to ask patients suffering from postural headaches whether a predisposing event may have caused a disruption of weak points in the meninges when taking their medical histories. Some of the patients in our series had a history of engaging in exercises that required the Valsalva maneuver, such as lifting a heavy object, coughing, singing with handclapping, and jump roping.

Variable treatment options are available for patients with SIH. Conservative treatment consists of bed rest and oral analgesics, hydration, caffeine, mineralocorticoids and glucocorticoids. Invasive methods can be used to treat patients without substantial benefits from these measures. In the majority of these cases, autologous epidural blood patching is thought to be the treatment of choice. Intrathecal or epidural saline infusions[16] are also considerable options. If subsequent blood patches only result in transient effects and ultimately fail, surgical options should be considered. Direct surgical ligation of a leaking meningeal diverticulum, epidural packing using blood-soaked Gelfoam, muscle pledgets, or fibrin glue[3,16] are primary techniques used to seal the site of CSF outlet.

CONCLUSION

The presentation of SIH consists of positional headache, low CSF pressure, and diffuse pachymeningeal enhancement on MR imaging. However, not all SIH patients present with the classical diagnostic triad. In addition, autologous epidural blood patching is thought to be the treatment of choice in the variant type of SIH patients with confirmed site of CSF leakage.

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