Spontaneous Intracranial Hypotension Associated With Bilateral Chronic Subdural Hematomas
—Case Report—

Mineko MURAKAMI, Kentaro MORIKAWA, Akira MATSUNO, Kenshi KANEDA*, and Tadashi NAGASHIMA

Departments of Neurosurgery and *Neurology, Teikyo University Ichihara Hospital, Ichihara, Chiba

Abstract

A 34-year-old female presented with spontaneous intracranial hypotension (SIH) manifesting as severe postural headache and meningism. Magnetic resonance (MR) imaging with gadolinium showed diffuse pachymeningeal enhancement. She developed bilateral chronic subdural hematomas 4 weeks after the onset of the symptoms. MR imaging showed descent of the midline structures of the brain. The bilateral chronic subdural hematomas were surgically drained, with no remarkable pressure. Postoperative MR imaging showed complete resolution of the pachymeningeal enhancement and reelevation of the midline structures of the brain. SIH is an uncommon and probably unrecognized condition because of the usually benign course. However, this case emphasizes that SIH is not entirely benign. SIH should be considered if there is no identifiable risk for intracranial hemorrhage, particularly in young patients. Neurosurgical intervention for the treatment of the underlying cerebrospinal fluid leak may be required if SIH persists.

Key words: intracranial hypotension, chronic subdural hematoma, magnetic resonance imaging

Introduction

Spontaneous intracranial hypotension (SIH) is rare but is increasingly being recognized as a clinical syndrome. SIH was first described in 1938 as a cause of postural headache. In general, postural headache related to intracranial hypotension occurs following invasive procedures including lumbar puncture, ventriculoperitoneal shunt placement, or head injury in association with the leakage of cerebrospinal fluid (CSF). The diagnosis is usually obvious and the condition generally runs a benign course. However, postural headache may occasionally occur without an apparent precipitating cause, and may be accompanied by more severe symptoms including meningismus, nausea and vomiting, dizziness, vertigo, tinnitus, abducens nerve paresis, diplopia, photophobia, visual field deficits, and depression of consciousness.

The etiology of SIH is not clearly understood, but current evidence suggests that CSP leakage along the spinal axis is a cause. Low CSF opening pressure at lumbar puncture can usually confirm the diagnosis of SIH. On the other hand, magnetic resonance (MR) imaging can also demonstrate the characteristics of SIH, including a diffuse symmetrical enhancement of the pachymeninx with gadolinium, and a generalized descent of the midline structures of the brain. In addition, subdural hygroma is frequently observed and occasionally can develop into subdural hematomas, which may complicate the clinical course. Clinically, misdiagnosis of SIH at the early stage could lead to the development of more progressive clinical manifestations. Thus, recognition of the early symptoms of SIH is vital for treatment and could avoid intracranial complications.

Here, we describe a case of SIH with bilateral chronic subdural hematomas which required surgical treatment.

Case Report

A 34-year-old previously healthy female suddenly
developed severe stiffness and pain in her suboccipital and bifrontal regions while she was driving a car. The headache was accompanied by nausea and vomiting, which was worse in the standing or sitting position and was completely relieved when lying down. Valsalva maneuvers exacerbated the headache. The headache persisted for 4 days. She was then admitted to our hospital. She had had a slight fever and throat pain approximately a week before the onset of the headache, but no apparent history of head trauma, CSF rhinorrhea or otorrhea.

Neurological examination found only slight nuchal rigidity. Her body temperature was 37.5°C and blood examination showed a moderately elevated erythrocyte sedimentation rate of 42 mm/hr and a C-reactive protein (CRP) level of 1.1 mg/dl. Computed tomography (CT) showed mild cerebral edema with slit ventricles and effacement of cortical sulci (Fig. 1). Lumbar puncture in the lateral decubitus position revealed a very low CSF opening pressure, below 30 mmH₂O. CSF examination found white blood cell count of $7 \times 10^6$/l, protein level of 82 mg/dl, glucose level of 61 mg/dl, and Cl level of 126 mEq/l. Gram staining revealed no organisms, and cultures were negative. Brain and cervical spine MR imaging with and without contrast agent showed diffuse enhancement of the cranial and spinal dura (Fig. 2).

The initial diagnosis was viral meningitis. The patient was treated with intravenous hydration, bed rest, and occasional administration of oral analgesics when the headache was exacerbated. Her symptoms improved gradually and she was discharged home 2 weeks after admission.

She was readmitted to the hospital 2 weeks after discharge, complaining of the reappearance and aggravation of the postural headache accompanied by nausea and vomiting. Repeat brain MR imaging, performed 20 days after the first study, revealed...
bilateral convexity subdural hematomas with mass effect on the lateral ventricles. The smooth, diffuse enhancement of cranial dura remained intense after administration of gadolinium (Fig. 3 left). Sagittal MR imaging showed the cerebellar tonsils were displaced below the level of the foramen magnum, and thepons was displaced anteriorly and compressed against the clivus (Fig. 3 right). The thalamus and the optic chiasm were also displaced downward. Blood examination showed no coagulation abnormalities.

The bilateral subdural hematomas were identified as chronic hematomas and were surgically drained. The dark, changed liquid blood flowed out under low pressure through the outer membranes of the hematomas, indicating that intracranial pressure was lower than that of common chronic subdural hematoma. Biopsy of the dura mater and the outer membrane of the hematoma showed no evidence of inflammation or malignancy. Pneumocephalus was developed after the surgery, but her symptoms resolved completely 2 weeks after the surgery. Further examination of the cause of CSF leakage was not conducted and the patient was discharged home without neurological deficits 17 days after admission.

Follow-up CT performed 4 weeks after the surgery revealed no abnormalities. Five months after discharge, she remains free of headache, and MR imaging demonstrated reelevation of the midline structures of the brain to their normal position as well as complete resolution of the cranial dural enhancement and the chronic subdural hematomas (Fig. 4).

**Discussion**

Postural headache is the most common characteristic symptom of SIH. The cause may be caudal displacement of the pain-sensitive structures in the cranial vault when the patient is upright, or that intracranial hypotension induces the vasodilation of the pain-sensitive blood vessels. Maneuvers such as jugular venous compression or Valsalva maneuver, which increase intracranial pressure by increasing intracranial blood volume, can exacerbate the headache even when the patient is supine. Nonpositional and persistent headache also occurs in patients with associated subdural effusion or chronic stage of the illness, but the headache at the onset of symptoms, either acute or gradual, is generally orthostatic.

The diagnosis is usually confirmed by a low CSF opening pressure on lumbar puncture of 60 mmH2O or less. CSF pressures were consistently over 90 mmH2O and as high as 130 mmH2O in three of 26 patients. Variable CSF pressure may indicate that the CSF leak is intermittent.

Abnormalities in CSF analysis are common in SIH, and include xanthochromia, lymphocytosis, and elevated protein level. These abnormalities may be due to the caudal displacement of the brain, which results in tearing of the meningeal blood vessels, or diapedesis of cells and protein into the subarachnoid space from the compensatory dilation of meningeal blood vessels. The finding of lymphocytosis may lead to an erroneous diagnosis of meningeal infection, particularly in the presence of meningismus and fever, as in our patient. However, fever is not a feature of SIH. In our patient, the fever, elevated erythrocyte sedimentation rate and CRP level were confirmed to result from upper respiratory tract infection.

MR imaging has recently been emphasized as the major method for the confirmation of the diagnosis. The MR imaging characteristics of SIH include diffuse symmetrical enhancement of the pachymeninx with gadolinium; generalized descent of the midline structures of the brain including optic chiasm, brainstem, and cerebellar tonsils which frequently resembles tonsillar herniation or Chiari type I malformation; effacement or crowding of the sulci, gyri, sylvian fissure, basal cisterns, and slit ventricles; enlarged dural sinuses; and enlargement of the pituitary gland. Diffuse dural enhancement is the most striking and valuable finding for the diagnosis. Other findings such as descent of the brain or effacement of the sulci and basal cisterns are not

Neural Med Chir (Tokyo) 40, September, 2000
easy to recognize in the early stage of the illness. The dural enhancement is presumably the result of either tearing or vasodilation of the meningeal vasculature, which lacks tight junctions.6,11,12 Similar meningeal enhancement may be found in infectious, granulomatous, and carcinomatous meningitis.6,11,12 However, in these diseases, the dural enhancement is usually patchy and often involves the leptomeninges in the basal cisterns. In SIH, the meningeal enhancement is diffuse, thick, continuous, and prominent in the cranial convexity, falx, and tent. Leptomeningeal enhancement, such as extension of enhancement to the cortical sulci or enhancement of leptomeninges around the brainstem, is rarely observed in SIH.6,11,12

The most important intracranial complications of SIH are subdural hematoma or hygroma and downward displacement of the cerebellar tonsils.1,3,5,6,10,16,18,20,23,28 Subdural hematoma may occur as a result of tearing of bridging veins in the layer of the dural border cells, and could develop into chronic subdural hematoma.6,8,11,13,16 Subdural hygroma or hematoma which is limited in volume will usually resolve spontaneously after normal CSF volume has been restored. However, a significant volume of hematoma may require surgical treatment. Two patients with SIH who developed subdural hematomas after intracranial pressure had normalized, presumably because of persistent fragility and dilation of the small dural vessels which resulted in vascular rupture and the delayed subdural hematomas.16 Caudal displacement of the cerebellar tonsils may be misdiagnosed as tonsillar herniation with intracranial hypertension in the acute stage of the illness, or idiopathic Chiari type I malformation in the chronic stage of the illness.3,4,13 Panpachymeningeal enhancement on MR imaging with contrast medium is important to distinguish SIH from these conditions. In addition, no clinical or radiographic evidence of syringomyelia was demonstrated in seven cases of acquired Chiari type I malformation secondary to SIH.3,10

CSF leakage along the spinal axis is increasingly recognized as the cause of SIH, and more attention has been paid to identifying such leakage.3,10,13,16,20,23 A small tear in the nerve root sleeves or spinal meningeal diverticula (Tarlov’s cyst) has been identified as a source of occult leakage, which may be anywhere within the spinal canal, but most commonly in the cervicothoracic region.15,16,20,22 Rupture of the meningeal diverticula or nerve root sleeves may occur even from minor stresses, such as a trivial fall, vigorous exercise, or violent coughing, and in some cases, local back pain at the level of the CSF leak precedes the onset of the headache.10,16,20,22

The etiology of meningeal diverticula is largely unknown, but some inherited connective tissue disorders are associated with spinal meningeal diverticula, including Marfan’s syndrome, Ehlers-Danlos syndrome, or neurofibromatosis.10,20,22 A patient with SIH due to occult CSF leakage who had hypothyroidism after the resection of a pituitary microadenoma indicates that hypothyroidism could predispose patients to the rupture of congenital meningeal diverticula or the formation of acquired diverticula.16 In our patient, we did not conduct any further investigation, such as radioisotope cisternography or CT myelography, to identify the site of the CSF leakage because the condition resolved spontaneously and no generalized connective tissue disease or hypothyroidism was found.

SIH usually resolves spontaneously in several weeks with conservative treatment such as bed rest and oral or intravenous hydration. Medical treatment consists of analgesics, sedatives, antiemetics, caffeine, or corticosteroid administration.4,13,16,20 The epidural blood patch is a more direct method of treatment.4,7,8,10,11,14,20,27 Continuous intrathecal or epidural infusion of saline is very useful in some cases.1,13,14,16,26 If conservative treatment fails and the site of CSF leakage has been identified, neurosurgical intervention may be required.

References


Address reprint requests to: M. Murakami, M.D., Department of Neurosurgery, Teikyo University Ichihara Hospital, 3426-3 Anegasaki, Ichihara, Chiba 299-0111, Japan.