SPONTANEOUS INTRACRANIAL HYPOTENSION : A CASE REPORT

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Spontaneous intracranial hypotension (SIH) is uncommon and can be misdiagnosed as Chiari I malformation, carcinomatous meningitis, idiopathic hypertrophic pachymeningitis, or subdural hematoma. If this is not recognized, it could lead to improper management. Many have been reported in the literature abroad, but to our knowledge, not in Thailand. Therefore we would like to present our case of SIH along with the overseas literature reviews.

CASE REPORT

A 42-year-old man presented with 1-month history of headache, predominantly at the occiput and a flashing pain along the right arm when he turned his neck. Previously, he had been healthy and had had no history of operation, trauma or lumbar puncture. Physical examination revealed no abnormality. Plain films of the cervical spine showed no abnormality.

MRI of the spine and brain was performed. Cranial sagittal and axial MR images revealed bilateral subdural effusions, generalized descent of midline structures that crowded the posterior fossa with tonsilar herniation, flattening of the pons against the clivus and elongated shape of the midbrain (Figure1-3). Postgadolinium enhanced images showed diffuse symmetrical pachymeningeal enhancement which is often referred to as pachymeningitis (Figure4). On MRI of the whole spine, there was only mild anterior epidural enhancement at the level of C2, but no spinal hygroma (subdural or epidural fluid collection) was observed.

The MRI findings are compatible with spontaneous intracranial hypotension. Additional information that the headache was relieved by lying down and that onset occurred after trying to move a heavy cabinet was obtained.

Conservative treatment was performed (high fluid intake and bed rest) and followed by clinical improvement.

Repeated cranial MR images obtained 4 months later demonstrated complete resolution of the disease. (Figure 5-6)

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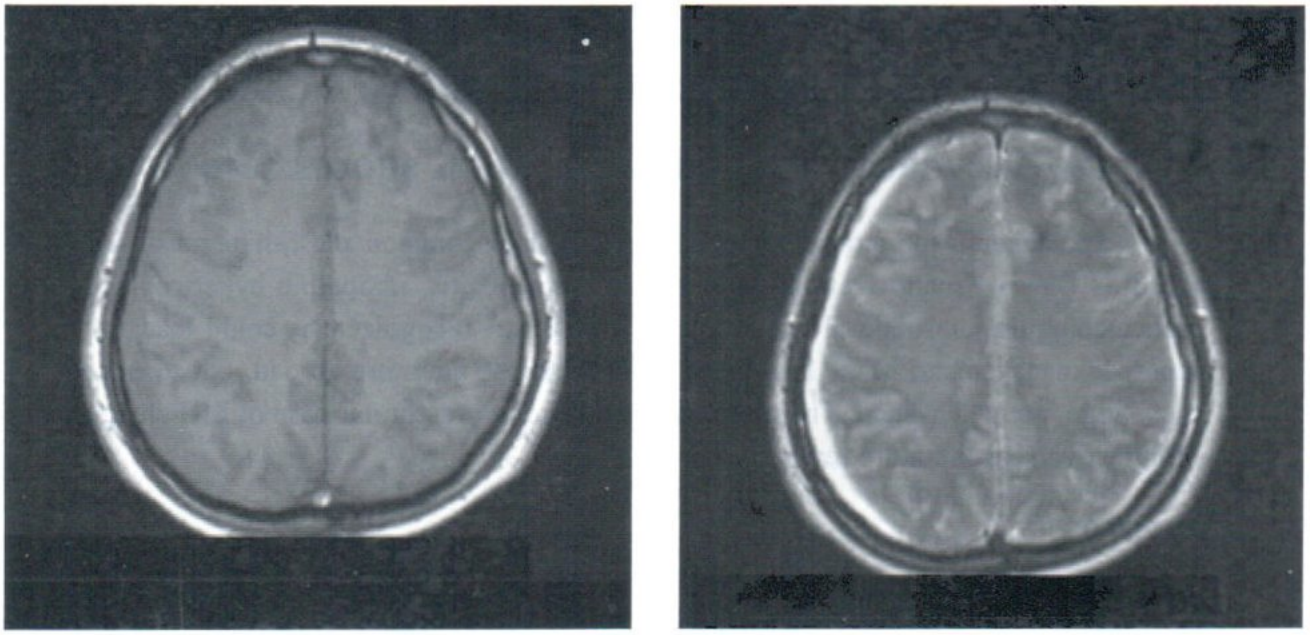
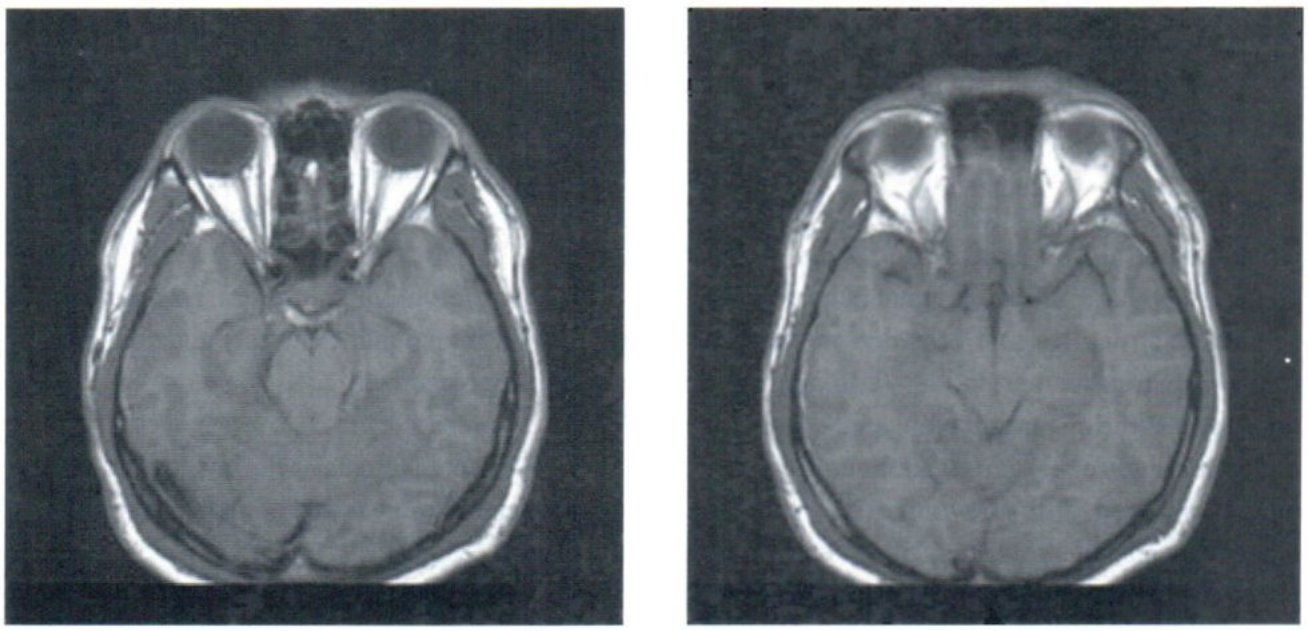


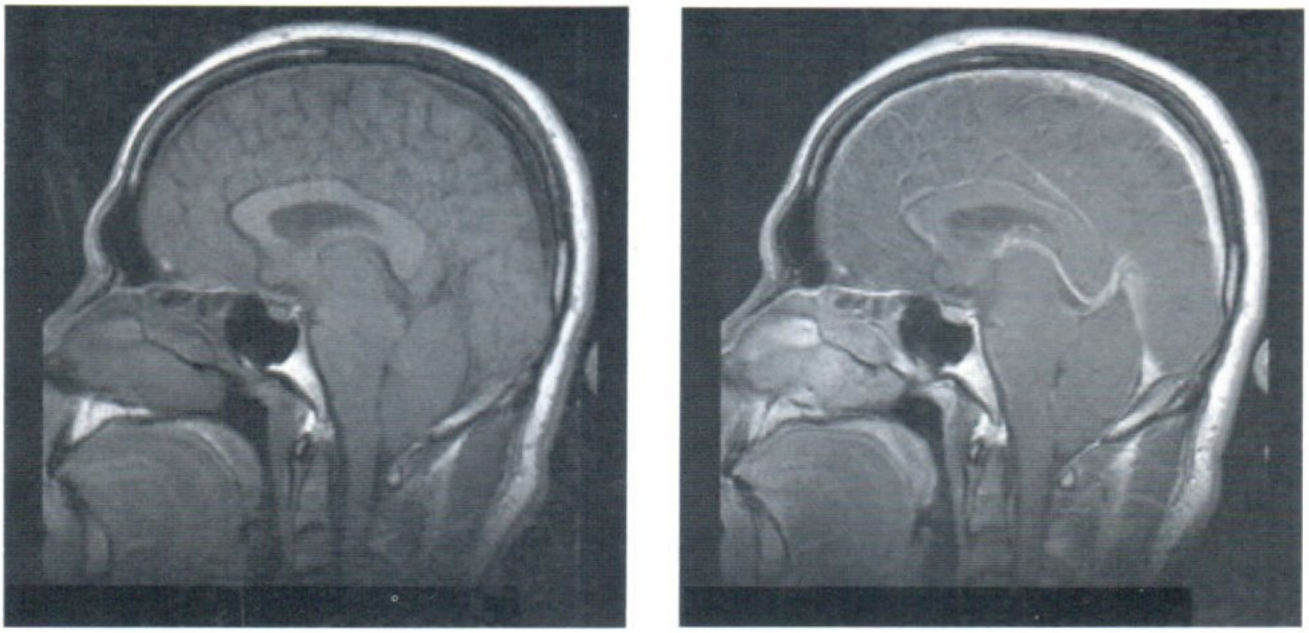
1A 1B

Fig. 1. Axial SE T1WI (A) and FSE T2WI (B) show bilateral subdural effusion.



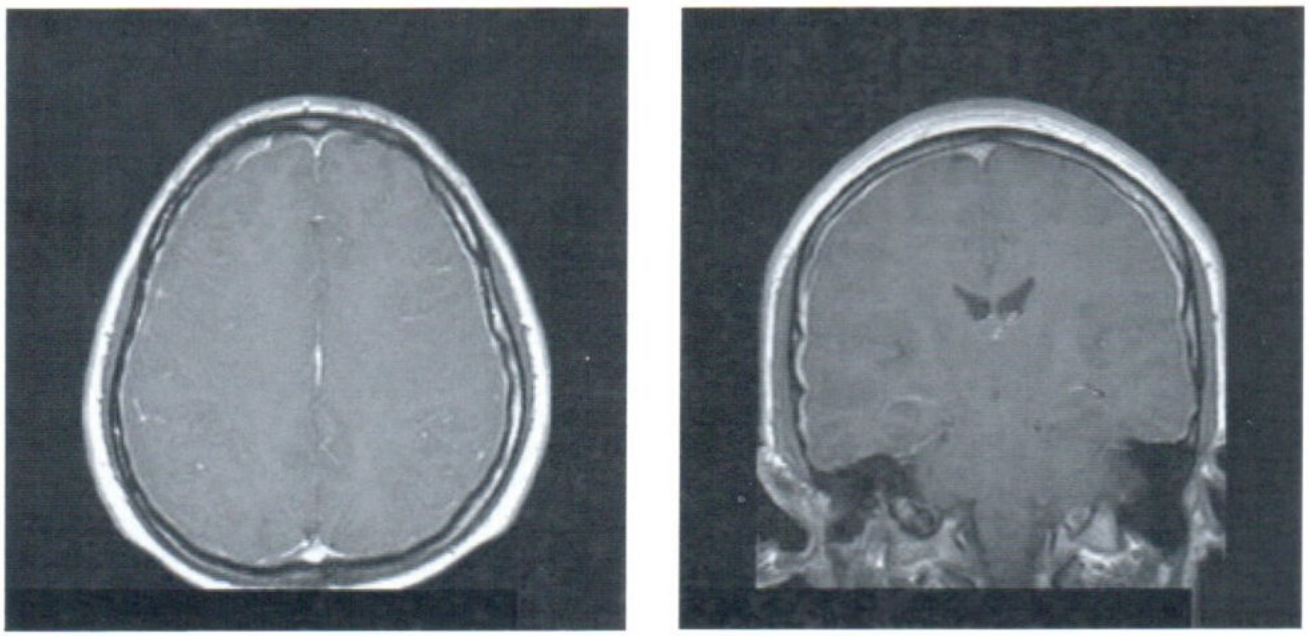
2A 2B

Fig. 2. Axial SE T1-weighted images at the level of midbrain show elongated shape of the midbrain in AP dimension with effacement of basal cisterns due to descent of the midline structure.



3A 3B

Fig. 3. Pregadolium-enhanced (A), and postgadolinium-enhanced (B) sagittal SE T1WI show flattening of the pons against the clivus and tonsilar herniation. Venous dilatation is also noted on the postcontrast image.



4A 4B

Fig. 4. Postgadolium-enhanced axial (A) and coronal (B) SE T1WI show diffuse symmetrical pachymeningeal enhancement.

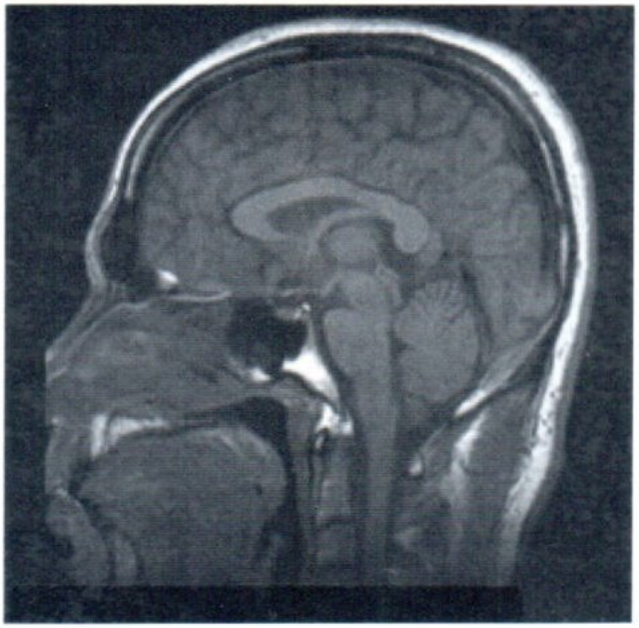
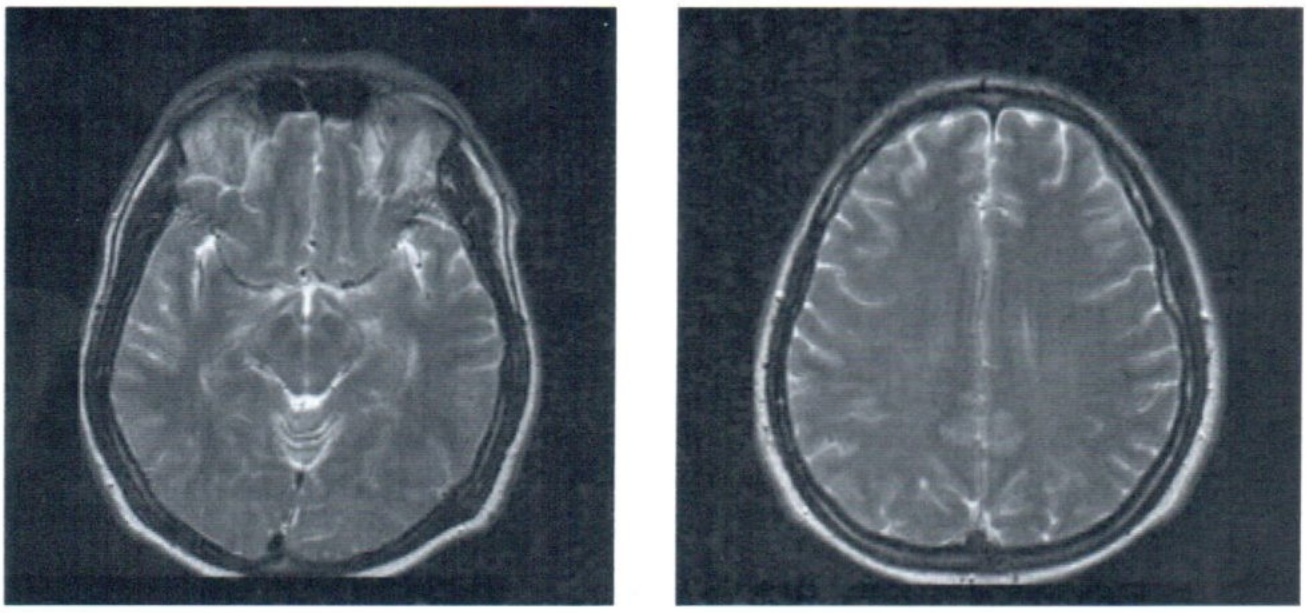


Fig. 5. Sagittal SE T1WI after conservative treatment demonstrates disappearance of flattening of the pons against the clivus and inferior displacement of the brain.



3A **3B**

Fig. 6. Axial FSE T2WI after conservative treatment 4 months later, shows normal appearance of the midbrain and basal cistern (A) and complete disappearance of the bilateral subdural collections (B).

DISCUSSION AND REVIEW OF LITERATURE

Spontaneous intracranial hypotension was first described by Schaltenbrand in 1938. Three mechanisms were suggested by Schaltenbrand: occult dural tear, overabsorption of CSF, and decreased CSF production. The similarity of SIH to the post-LP headache syndrome supports the notion that a CSF leak may be the cause.

In 1968, Teng and Papatheodorou summarized findings in 31 cases and found a female preponderance of 3.4:1 with the majority of patients in the third or fourth decade of life.

Epidemiological studies have not been reported, but the prevalence of SIH in Olmsted County, Minnesota, in 1995 was approximately one in 50,000 patients (unpublished data).[18]

The leak is typically at the level of the spine, particularly the thoracic spine and cervicothoracic spine, rarely at the skull base. CT myelography and radionuclide cisternography are the most useful radiographic studies to diagnose and localize the spinal CSF leak.

The cause of CSF leakage reported in the literature are a cervical bone spur,[14] disc herniation,[2] meningeal diverticula, tear in the nerve root sleeves, or Tarlov's cyst. However it often remains undetermined. In a substantial minority of patients, there is a history of trivial or minor trauma (as in our case)or evidence of connective tissue disease (eg. Marfan's syndrome, Ehlers-Danlos syndrome, NF, ADPCK, etc)[4,5]

The common clinical features of SIH are orthostatic headaches, neck or interscapular pain, nausea, emesis, horizontal diplopia, change in hearing, blurring of vision, facial numbness and upper limb radicular symptoms.[3,6,15,16]

The headache associated with SIH is probably caused by dilatation of cerebral veins and meningeal vasculature because there is a reciprocal relationship between CSF volume and intracranial blood volume (The Monroe-Kelly rule)[9]; it may be a consequence of the low CSF pressure producing displacement of pain-sensitive structure (particularly traction of the intracranial dura); or both factors may be involved.[1]

The average human brain weights approximately 1400g in air. When floating freely in CSF, the human brain has an effective weight of approximately 50 g. The brain depends greatly on the antigravity effect of CSF to maintain its delicate structure. Robbed of this buoyancy, the brain may sag, collapsing in on itself.[15]

The headache may be gradual or acute in onset and may be generalized or localized to the frontal or occipital regions.[10]

Visual disturbances are related to occur in 23% of patients and may be attributed to distortion of the optic chiasm or compression or vascular congestion of the intracranial portions of the optic nerves.

Auditory or vestibular symptoms include hypoacusis, hyperacusis, tinnitus, nausea, vomiting and dizziness. These are reported to occur in approximately 20% of cases and probably result from a change in intralabyrinthine pressure transmitted through the cochlear aqueduct.[1]

The diagnosis of intracranial hypotension is confirmed by a low (< 6cm H2O) or low-normal opening pressure on LP. Results of examination of CSF may be normal or show mildly raised

protein levels, increased cell count, or xanthochromia; these may be caused by meningeal hyperemia resulting from the low CSF pressure, accompanied by diapedesis of cells into the subarachnoid space or from disruption of the normal hydrostatic and osmotic pressure across the venous sinuses and arachnoid villi, accompanied by protein accumulation.[1,12]

On cranial MR imaging, subdural effusions are seen in approximately 10% of cases and are probably the result of the rupture of bridging veins caused by the decrease in CSF volume and downward displacement of the brain. Subdural effusions or hematomas in SIH have been shown not to be under pressure. Diffuse thickening of the meninges and choroid plexus, and meningeal enhancement may result from dilated meningeal vessels and small vessel rupture.[7]

In a few cases, meningeal biopsy samples have either shown nonspecific inflammation or no abnormality, supporting the hypothesis that dural enhancement is caused by venous enlargement.

The spinal manifestations associated with SIH-spinal dural enhancement, spinal epidural venous engorgement, subdural or epidural collections (spinal hygroma)- have been reported by several authors.[8,17]

Conservative treatment, with bed rest and high fluid intake, often suffices. Symptoms, however, can take several months to resolve. A short course of steroid medication or orally administered caffeine may be considered.[2]

If conservative measures fail to resolve symptoms, then extradural saline infusions or and epidural blood patch can be effective at stopping the leak, even if the source of the CSF fistula has not been demonstrated.[11] The epidural blood reportedly can travel up to nine spinal segments from its site of placement. Epidural blood produces an organized clot that could effectively tamponade any dural CSF leak. The rapid relief from headache seen immediately after the infusion of the blood must occur by some other mechanism, such as an increase in subarachnoid pressure. Placement of epidural blood patch can be repeated at the same level or applied directly at the site of CSF fistula.[11]

The indications for surgical treatment of spontaneous spinal CSF leakage have not been firmly established. The most straightforward indication may be the presence of persistent symptoms in spite of the placement of multiple epidural blood patches in a patient in whom a discrete leaking meningeal diverticulum has been radiographically demonstrated. In addition, surgery may be considered as the primary treatment in all patients who have such meningeal diverticula, particularly when the patients are young and the diverticula are large. Spinal meningeal diverticula may grow and cause other neurological symptoms.[18]

In our case, history of postural related headache was initially missed. If we did not recognize this condition, it would have been misdiagnosed as bilateral chronic subdural hematomas, and proper management would have been delayed. Lumbar puncture was not done in this case, because the MR findings, symptom of postural headache and absence of history of LP were sufficient to diagnose SIH. The patient has been asymptomatic for 7 months up to now after conservative treatment. So further investigation for the site of CSF leakage (myelography or radionuclide cisternography) and epidural blood patching were not performed.

CONCLUSION

The imaging and clinical manifestation of

SIH have become well known, many patients have been diagnosed who otherwise would have been misdiagnosed with migraine, headache of unknown origin, aseptic meningitis, or subdural hematomas. Attention to the myriad manifestations of CSF hypovolemia both intracranially and extracranially will prevent such errors of diagnosis and facilitate prompt treatment of CSF fistula.

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