PONTANEOUS intracranial hypotension is a rare disease. Its presentation consists of severe positional headache and low CSF pressure associated with various neuroradiological findings such as leptomeningeal thickening and subdural hematoma on MR imaging and, occasionally, identification of leakage on RI cisternography or CT myelography. Spontaneous intracranial hypotension can be induced by a trivial fall, vigorous exercise, or a violent bout of coughing. Because of the difficulty frequently encountered in demonstrating the site of leakage, other causative factors such as reduced CSF production or hyperabsorption of CSF have been proposed by some authors. However, it is currently accepted that the major causative factor of SIH is the release of CSF due to an underlying weakness of the spinal meninges, even though no leakage may be demonstrated. In the present case, we found direct evidence of CSF leakage using MR myelography, although other diagnostic methods failed to demonstrate definitive findings. The usefulness of MR myelography and the pathogenesis of SIH are discussed.

Case Report

This 26-year-old woman was referred to our hospital with the complaints of sudden headache and vomiting following straining. She stated that the headache worsened when she moved into a standing position.

Examination. Lumbar puncture revealed an initial pressure of 0 cm H2O as well as a cell count of 42/3 mm3 (predominantly composed of monocytes), 115 mg/dl protein, 50 mg/dl glucose, and 123 mEq/L chlorine. One month after onset of symptoms, brain MR imaging revealed right-sided subdural effusion and diffuse thickening of the leptomeninges. The disease was diagnosed as SIH syndrome.

Conventional myelography revealed no sign of CSF leakage (Fig. 2 left). An 111 In-DTPA (the diethylenetriamine pentaacetic acid chelate of indium 111) scintigraphy study performed 3 hours after injection of the RI also revealed no abnormal leakage; however, there was rapid washout of indium-111 from the CSF space (Fig. 2 right). Computerized tomography myelography, which was performed following conventional myelography, demonstrated thickening of the epidural space and thickened pleura in the right dorsal thoracic cavity (Fig. 3). Delayed CT myelography performed 24 hours after injection of contrast agent revealed disappearance of the intradural contrast agent, but no findings of leakage.

Thoracic MR imaging was performed using a highly T2-weighted sequence (TR 5082 msec, TE 240 msec)—so-called MR myelography. A coronal image revealed a high-intensity stripe originating in the vicinity of the T4–5 level and extending obliquely downward (Fig. 4). Conventional T2-weighted MR imaging did not demonstrate

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computerized tomography; RI = radioisotope; SIH = spontaneous intracranial hypotension.
clear evidence of the leakage site, but there was dural thickening on the axial images.

**Treatment and Outcome.** The patient’s symptoms recovered in response to a period of bed rest lasting approximately 2 months. The high-intensity stripe observed on MR myelography also disappeared 5 months after symptom onset (Fig. 5).

**Discussion**

Spontaneous intracranial hypotension is a rare but increasingly recognized pathological entity. Its clinical features are characterized by postural headache and low CSF pressure. These are often associated with intracranial dural thickening, subdural hematoma, and downward displacement of the brain. Radioisotope cisternography findings are characterized by rapid clearance of the RI from the CSF space, early appearance of urinary bladder activity, and identification of the site of the leakage in some cases. Possible causes of this disease are thought to be diminished CSF production, hyperabsorption of CSF, or CSF leakage. Among these causes, CSF leakage due to an underlying weakness of the spinal meninges is currently recognized as a major cause, although the leakage site cannot always be demonstrated.

In our case, RI cisternography demonstrated a rapid washout of the RI from the CSF space, but no abnormal leakage. The sensitivity of RI cisternography is not very high (60%) and, moreover, the spatial resolution of this technique is poor. Conventional myelography and CT myelography also could not detect the source of leakage. In some cases, leakage can be demonstrated by CT myelography. However, it is difficult to examine a wide area using axial CT scanning if the leakage site is unclear.

Magnetic resonance myelography (cisternography) can enhance the CSF signal by suppression of the adjacent tissue’s signal. Magnetic resonance myelography is also known to possess a superior capability to detect a CSF fistula of the skull compared with CT cisternography. There is a single case description in which an axial T$_2$-weighted image depicted CSF leakage along the course of a nerve root. However, using the conventional spin-echo sequence for T$_2$-weighted imaging, MR imaging does not suppress the signal of adjacent tissues and the detectability of the site of a fistula is low. Epidural fatty tissue...
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appears bright when the spin-echo type of T2-weighted imaging is used. Magnetic resonance myelography eliminates the signal from epidural fatty tissue due to the technique’s extremely long echo time and effective fat suppression.

The use of MR myelography for detecting leakage of CSF in cases of SIH has not appeared previously in the literature. In our case, MR myelography demonstrated a leak that traveled along the intercostal nerve and made an oblique descent that, based on the fact that there was no pleural effusion on chest x-ray films, probably led to the extrapleural space. However, there was thickening of the pleura in the right dorsal thoracic cavity, which was demonstrated on CT scanning. From these findings, it may be assumed that CSF leaked from the intercostal perineural space into the extrapleural space.

The rapid clearance of RI in cases of SIH may be explained by the theory that the RI leaks from the CSF space and spreads into the extrapleural space. It remains unclear why there was no leakage shown on RI cisternography in our case. The detection sensitivity of RI cisternography is approximately 60% in cases of SIH; thus nondiagnostic results exist in nearly half of the cases. We may speculate that the leakage into the extrapleural space was below the detectability of RI cisternography or there was no active CSF leakage at the time of the examination. Computerized tomography cisternography and/or RI cisternography studies are known to have higher rates of detectability when active CSF leakage is present at the time of the diagnostic procedure or, even better, during the Valsalva maneuver. On the contrary, because MR myelography is capable of detecting the CSF itself, MR myelography may detect the pool of leaked CSF and not necessarily active leakage of contrast media or RI. Schaltenbrand initially believed that the cause of SIH was CSF hyperabsorption, based on the rapid clearance of CSF demonstrated on RI scintigraphy. However, based on the CSF leakage theory and findings in the present case, we assume that the early clearance of RI is due to rapid absorption from the leakage site, that is, the spinal epidural space and/or the extrapleural space as shown in our case.

El Gammal and colleagues stated that an accurate diagnosis of an intracranial CSF fistula cannot be made unless the high-intensity signal displayed by the fistulous communication is observed to be directly continuous with the intracranial subarachnoid space. In our case, there was no clearly defined communication of leakage with the dural sac. This may be due to a motion artifact and/or a very narrow primary leakage site. With an improved acquisition technique and accumulation of similar cases, this question may be solved in the future. To delineate the exact site of leakage more clearly, we should have performed axial MR myelography after obtaining the coronal slice along the spinal axis. However, if one only has the option of using CT myelography to find the fistula, it is difficult and time consuming to cover the entire spinal axis with axial slices when no indication of leakage site is evident. Therefore, we suggest that coronal MR myelography be performed first, followed by axial MR imaging and/or focused CT myelography, if necessary.

Although our report contains only a single case description, we emphasize that MR myelography may increase the physician’s diagnostic ability to clarify the cause and site of CSF leakage in cases in which the origin of SIH is
unknown. Magnetic resonance myelography is a noninvasive method that is highly sensitive in detecting water in tissue and, therefore, this method should be used routinely in the diagnosis of SIH.

References

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