Cranial MRI predicts outcome of spontaneous intracranial hypotension

Abstract—The outcome of spontaneous intracranial hypotension has been unpredictable. The results of initial MRI were correlated to outcome of treatment in 33 patients with spontaneous intracranial hypotension. A good outcome was obtained in 25 (97%) of 26 patients with an abnormal MRI vs only 1 (14%) of 7 patients with a normal MRI ($p = 0.00004$). These findings show that normal initial MRI is predictive of poor outcome in spontaneous intracranial hypotension.

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Spontaneous intracranial hypotension is increasingly recognized as an important cause of new daily persistent headaches, although an initial misdiagnosis remains common.¹ The cause of spontaneous intracranial hypotension is a spontaneous spinal CSF leak often associated with an underlying generalized connective tissue disorder.² Most cases of spontaneous intracranial hypotension are believed to be self-limiting, and initial treatment is centered around a course of bedrest and hydration. Nevertheless, persistent symptoms are present in the majority of patients who come to medical attention; for those, a variety of treatment options are available, including epidural blood patching,³ percutaneous fibrin sealant placement,⁴ and surgical repair of the underlying CSF leak.⁵ The reported results of these various treatments have generally been good, but the outcome of spontaneous intracranial hypotension is unpredictable and some patients have persistent and often incapacitating symptoms in spite of maximal medical and surgical treatments. The vast majority of patients with spontaneous intracranial hypotension undergo cranial MRI scanning early in their clinical course prior to any therapeutic intervention. It has been our experience that patients with recalcitrant symptoms generally have had normal MRI findings. We therefore investigated a large group of patients with spontaneous intracranial hypotension to determine whether abnormalities on initial MRI can predict outcome.

Methods. The patient population consisted of a group of 33 consecutive patients with spontaneous spinal CSF leaks and intracranial hypotension who were referred to us for evaluation and treatment. The mean age of the 23 women and 10 men was 41 years (range 13 to 72 years). The presenting symptom was a positional headache in 31 patients, a nonpositional headache in one patient, and nonpositional neck pain in one patient. Cranial MRI was available for all patients, which was reviewed for features of intracranial hypotension. The presence of a spinal CSF leak was confirmed by CT myelography in all patients. Radionu-

clide cisternography was performed in only a few patients because CT myelography has almost completely replaced this nuclear medicine study in our practice. None of the patients had a cranial CSF leak. Treatment consisted of 1) conservative measures such as bedrest, oral hydration, oral caffeine, and use of an abdominal binder; 2) high-volume epidural blood patching (up to 80 mL) injected at the thoracolumbar junction; 3) percutaneous placement of a fibrin sealant; or 4) surgical repair of the CSF leak. Follow-up was complete for all patients with a minimum duration of follow-up of 3 months from the time of last treatment. A good outcome was defined as complete or near-complete resolution of symptoms and a poor outcome as no or only minimal improvement with a continued search for effective treatments. The Fisher exact test was used for statistical analysis.

Results. Clinical and radiographic features of the patient population are summarized in the table. Twenty-six (78%) of the 33 patients had an abnormal initial MRI, and 7 (22%) had a normal initial MRI. MRI abnormalities included pachymeningeal enhancement in 24 patients, downward displacement (sagging) of the brain in 17 patients, and subdural fluid collections in 14 patients. A good outcome was obtained in 25 (97%) of 26 patients with an abnormal MRI vs only 1 (14%) of 7 patients with a normal MRI ($p = 0.00004$). Patients whose initial MRI was normal never developed any abnormalities on follow-up MRI studies. All patients with a normal initial MRI had multiple spinal CSF leaks. These patients underwent multiple therapeutic interventions including surgeries with CT myelographic confirmation of successful repair of all visualized CSF leaks (figure). Diagnostic intrathecal saline infusions were performed in five of the six patients with normal MRI who had persistent symptoms. Positional symptoms resolved in four of these five patients during saline infusion, suggesting the presence of an ongoing CSF leak in the setting of a normal CT myelogram.

Discussion. In this study, initial MRI findings were highly predictive of the outcome of spontaneous intracranial hypotension. Patients with an abnormal MRI had an almost universally good outcome, whereas those with a normal MRI generally had a poor outcome. The reason for this dichotomy is unclear.

MRI has had a major impact on the recognition and our understanding of spontaneous intracranial hypotension.⁶ Prior to MRI, lumbar puncture was an essential part of the diagnostic evaluation, and the low opening pressure had to be recognized as abnormal. Moreover, normal opening pressures are not uncommon among patients with spontaneous intracranial hypotension. The typical MRI findings of in-

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Table. Characteristic of 33 patients with spontaneous intracranial hypotension

	Cranial MRI	
	Abnormal	Normal
No. of cases	26	7
M/F	9/17	1/6
Age, mean (range); y	42 (22–72)	37 (13–45)
Positional headache	24	7
Time to first MRI, mean (range)	4 wk (2 d to 7 mo)	3 wk (1 d to 2 mo)
Time to diagnosis, mean (range)	34 wk (2 d to 13 y)	22 wk (1 wk to 2 y)
Type of CSF leak		
Single	7	0
Multiple	19	7
Site of CSF leak		
Cervical	16	7
Thoracic	15	6
Lumbar	2	0
Treatment		
Conservative only	2	0
Epidural blood patch	24	7
Percutaneous fibrin sealant	6	3
Surgery	13	7

tracranial hypotension are more easily recognized and include pachymeningeal enhancement, subdural fluid collections, prominence of cerebral venous sinuses, and pituitary hyperemia.^{1,6,7} These abnormalities are believed to be compensatory mechanisms related to the loss of CSF volume.^{7,8} Another frequent MRI finding is downward displacement of the brain, or brain sagging, which is a result of the loss of CSF buoyancy.^{1,6} The meningeal enhancement, in particular, has in the past been considered to be the sine qua non of intracranial hypotension,⁶ but it has become apparent that meningeal enhancement—or any of the other features of intracranial hypotension—may be absent in patients with spontaneous intracranial hypotension.^{9,10} In our patient population, about one-fifth of patients had normal MRI. It should be noted, however, that our patient population is skewed toward difficult-to-treat patients seeking surgical repair of their CSF leaks. The diagnosis of spontaneous intracranial hypotension in our patient population is incontrovertible because all patients had clear evidence for an underlying spinal CSF leak on CT myelography with surgical confirmation.

One possible explanation for the findings in our study would be that patients with normal MRI scans are more difficult to diagnose and thus there is a longer delay until effective treatment can be initiated. However, the time from onset of symptoms to first MRI and to diagnosis was very similar among

Figure. Imaging in spontaneous intracranial hypotension. (A) Coronal enhanced T1-weighted MRI shows typical changes of intracranial hypotension with subdural fluid collections (arrows) and meningeal enhancement (arrowheads) in a 35-year-old man with multiple spontaneous spinal CSF leaks. (B) Normal coronal enhanced T1-weighted MRI in a 43-year-old man with spontaneous CSF leaks at the T2 to T3 (C, arrow) and T6 to T7 (D, arrows) levels on CT myelography. (E, F) Following surgical repair, no CSF leak was detected, but symptoms persisted. An arachnoid cyst arising from the left T7 nerve root was ligated with an aneurysm clip (F, arrow).

the two groups of patients. Another possible explanation would be that the spinal CSF leaks in patients with normal MRI are different and more difficult to treat. However, the site, multiplicity, and radiographic appearance of the CSF leaks on CT myelography were very similar among the two groups of patients.

We postulate that patients with spontaneous intracranial hypotension and normal MRI lack the ability to compensate for any loss of CSF volume and thus continue to experience symptoms when very small CSF leaks persist in spite of maximal medical and surgical treatments.

Further investigations in larger patient populations are required to confirm or refute the findings in our

study that a normal initial MRI is predictive of a poor outcome in spontaneous intracranial hypotension.

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Central paroxysmal positional vertigo

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A 75-year-old hypertensive man had episodic vertigo when lying down or getting up. There was no ophthalmoplegia, saccadic pursuit, spontaneous or gaze-evoked nystagmus, weakness, or cerebellar ataxia. In the supine position, vertigo and a left-beating purely torsional nystagmus appeared a few seconds after the head was turned to the left. Nystagmus persisted for approximately 1 minute while the head was maintained in that position but fatigued after two repetitions of the position. Treatment with the Epley maneuver was unsuccessful. MRI showed a small infarct located dorsolateral to the fourth ventricle (figure), which suggests that interruption of the vestibular nuclei–archicerebellar loop seems to be responsible for central paroxysmal positional vertigo.¹

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Figure. Diffusion-weighted axial MRI shows an acute infarct located dorsolateral to the fourth ventricle (arrow). The vestibulocerebellar and fastigiovestibular fibers are likely to be involved in the lesion. Magnetic resonance angiograms of the posterior circulation were unremarkable.

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