Expert Opinion

Spontaneous Intracranial Hypotension Resulting in Coma

Case History Submitted by Randolph W. Evans, MD
Expert Opinion by Bahram Mokri, MD

Key words: spontaneous intracranial hypotension, coma
(*Headache.* 2002;42:159-160)

This case expands our knowledge of the spectrum of manifestations of spontaneous intracranial hypotension.

CLINICAL HISTORY

A 42-year-old man was seen for a third neurological opinion for obtundation. He presented with a 1-month history of new-onset, daily, intense, right-sided headaches (information about postural precipitation is not available). A magnetic resonance imaging (MRI) scan of the brain showed diffuse meningeal enhancement. A cervical MRI showed only bulging disks. A lumbar puncture 2 days after admission to the hospital was performed without measurement of the opening pressure. The cerebrospinal fluid (CSF) revealed a glucose of 47 mg/dL, a protein of 112 mg/dL with 10 white cells (5 lymphocytes), and 650 red blood cells. The CSF cryptococcal antigen and cultures were negative. Testing for human immunodeficiency virus (HIV) was negative. A cerebral arteriogram was negative. A meningeal biopsy showed acute and chronic inflammation but no evidence of neoplasm, sarcoid, or infection. A second lumbar puncture 10 days after the first produced an opening pressure too low to measure. There were 63 white cells (61% lymphocytes), a glucose of 58 mg/dL, and a protein of 88 mg/dL. The CSF Venereal Disease Research Laboratory (VDRL) test was nonreactive.

On admission he was alert, and the neurologic examination was normal. Over the next several days, he was intermittently lethargic. Over the following 5 days, he became increasingly lethargic and stopped following commands. He was intubated because of aspiration pneumonia, but the blood gases were satisfactory. When I saw him 4 days later, he was intubated, his eyes were closed, and he did not follow commands. Oculocephalics appeared intact. The pupils were 3 mm and reactive to light. He withdrew all extremities to painful stimuli. Plantars were extensor bilaterally.

A repeat MRI scan of the brain revealed small, bilateral, subdural fluid collections over the hemispheres, cerebellar tonsils below the foramen magnum, and decreased fluid in the suprasellar and chiasmatic cisterns. An MRI scan of the cervical, thoracic, and lumbar spine showed no evidence of a CSF leak. The initial MRI scan of the brain had been sent for another opinion to a neuroradiologist in San Francisco who conferred with Dr. Robert Fishman. Dr. Fishman suggested instillation of a large-volume, lumbar, epidural blood patch, and this subsequently was performed with 50 mL of autologous blood and the patient in the Trendelenburg position.

Within 1 day of the blood patch, the patient was awake but confused. Examination showed a pupil-sparing right third nerve palsy. An MRI scan of the brain 3 days after placement of the blood patch had demonstrated some restoration of CSF in the chias-

Address correspondence to Dr. Randolph W. Evans, Suite 1370, 1200 Binz, Houston, TX 77004 or Dr. Bahram Mokri, Department of Neurology, Mayo Clinic, 200 First Street SW, Rochester, MN 55905.

matic and suprasellar cisterns and elevation of the brain stem. Two weeks after the blood patch, he was alert and oriented and had no recollection of the prior month. On follow-up 6 months later, he was doing well except for a resolving third nerve palsy.

Question.—Could this be a case of spontaneous intracranial hypotension?

EXPERT COMMENTARY

In the past decade, recognition of the MRI abnormalities characteristic of spontaneous intracranial hypotension (SIH) has revolutionized our understanding of this syndrome. Many physicians heavily involved in diagnosis and management of patients with headache are seeing many more patients with SIH than before.

It appears that most, if not all, cases of SIH result from spontaneous CSF leaks. These spontaneous leaks are typically at the level of the spine, particularly the thoracic spine and cervicothoracic junction, and only rarely at the level of the skull base. The CSF opening pressure is typically but not invariably low. The CSF analysis sometimes may yield entirely normal results, but more commonly shows an increased protein concentration and a white cell (primarily lymphocytic) pleocytosis, at least at some point during the course of the syndrome. Although these evaluations of protein and white cell count occasionally can be alarmingly high (protein concentration of 300 or higher and pleocytosis in the 200s), the CSF sugar is never low, and CSF cytology and microbiology are always negative.[1]

In its classic form, the syndrome is manifested by orthostatic headaches, low CSF pressure, and diffuse pachymeningeal gadolinium enhancement by contrasted brain MRI. The variability, however, is considerable. Some patients with typical clinical manifestations, very low CSF pressures, and documented CSF leak may not show pachymeningeal enhancement on MRI, while some other patients with typical clinical and imaging features of the syndrome and documented CSF leaks may display CSF opening pressures that are consistently normal. Yet another group of patients with typical imaging features of the syndrome, low CSF pressures, and documented CSF leaks or shunt overdrainage may lack headaches. In addition to these four major presentations ("classic" form, "normal meninges" form, "normal pressure" form, and "acephalgic" form),[1] isolated cases with unusual presentations have been reported; these have included a patient with parkinsonism, ataxia, and bulbar weakness,[2] a patient with severe encephalopathy,[3] and a patient with stupor, presumably resulting from diencephalic compression.[4] There can be little doubt that further reports of unusual presentations will appear in the literature in the years to come.

The case presented here is another example of an unusual clinical presentation of this syndrome with the onset of severe bilateral headache followed by profound obtundation. The patient's brain MRIs reportedly demonstrated diffuse pachymeningeal enhancement, low-lying cerebellar tonsils, bilateral small subdural fluid collections, and a decrease in the size of perichiasmatic cisterns. The CSF opening pressure was very low, and the patient responded favorably to placement of an epidural blood patch. Despite the unusual clinical presentation, and although the actual site of the leak was not identified, the constellation of low CSF pressure, the abnormalities on brain MRI, and the clinical improvement following patch placement all clearly point to a diagnosis of SIH.

REFERENCES

1. Mokri B. Spontaneous cerebrospinal fluid leaks: from intracranial hypotension to cerebrospinal fluid hypovolemia—evolution of a concept. *Mayo Clin Proc.* 1999;74:1113-1123.
2. Pakiam AS, Lee C, Lang AE. Intracranial hypotension with parkinsonism, ataxia, and bulbar weakness. *Arch Neurol.* 1999;56:869-872.
3. Beck CE, Rizk NW, Kiger LT, Spencer D, Hill L, Adler JR. Intracranial hypotension presenting with severe encephalopathy. *J Neurosurg.* 1998;89:470-473.
4. Pleasure SJ, Abosch A, Friedman J, et al. Spontaneous intracranial hypotension resulting in stupor caused by diencephalic compression. *Neurology.* 1998;50:1854-1857.