Intracranial hypotension presenting with severe encephalopathy

Case report

CARTER E. BECK, M.D., NORMAN W. RIZK, M.D., LYDIA T. KIGER, M.D., DAVID SPENCER, M.D., LAUREEN HILL, M.D., AND JOHN R. ADLER, M.D.

Departments of Neurosurgery and Neurology, and Division of Pulmonary and Critical Care Medicine, Stanford University School of Medicine, Stanford, California

A patient with severe and protracted symptoms from intracranial hypotension is described. The patient's presentation was marked by diffuse encephalopathy and profound depression of consciousness. This case report expands the presently known clinical spectrum of this uncommon and generally benign illness. The clinical and laboratory findings typically observed in the syndrome of intracranial hypotension are outlined. The pathophysiological mechanisms of the phenomenon are briefly discussed. Intracranial hypotension is a potentially severe illness with specific treatments that are distinct from the treatment of most neurological diseases. Three cardinal features—postural headache, pachymeningitis, and descent of midline cerebral structures—should prompt the diagnosis.

KEY WORDS • intracranial hypotension • encephalopathy • pachymeningitis • cerebrospinal fluid leak

Most patients with intracranial hypotension present with clinically mild symptoms, such as postural headache, that resolve spontaneously in only a few days. In its mildest form, intracranial hypotension is a common disorder that occurs following the performance of a significant proportion of procedures such as lumbar puncture and myelography. It may also complicate ventriculoperitoneal shunt placement in the so-called "overshunting syndrome." A growing literature provides description of a more severe syndrome whose manifestations include postural headache, meningism, nausea and vomiting, dizziness, cranial nerve palsies, blurred vision, diplopia, photophobia, and, rarely, seizures. Both spontaneous cerebrospinal fluid (CSF) leaks and iatrogenic CSF leaks have been reported as the cause of the syndrome. Because the source of leakage is often occult, intracranial hypotension frequently is not diagnosed in patients initially, which allows the illness to develop progressively more severe manifestations. The source of occult leakage has been identified in some cases as a dural tear or spinal meningeal diverticula (Tarlov's cyst). The syndrome of intracranial hypotension has been recently reviewed. Findings associated with this syndrome are briefly outlined below.

A characteristic constellation of findings is observed on magnetic resonance (MR) imaging. First, gadolinium-enhanced images show diffuse symmetrical pachymeningeal enhancement, which is often referred to as pachymeningitis. Axial images show effacement or crowding of the sulci, gyri, sylvian fissures, and basal cisterns. Mid-sagittal images demonstrate a generalized descent of midline structures that crowd the posterior fossa. The thalamus is displaced partly through the incisura, the cerebral aqueduct descends into the posterior fossa, the pons is deformed against the clivus, and there is tonsillar herniation (Fig. 1 left). Last, bilateral subdural hygromas frequently develop; they occasionally progress to form secondary hemorrhages.

As expected in intracranial hypotension, lumbar puncture is often notable for a low opening pressure, but not invariably. Cerebrospinal fluid analysis may be normal or show a lymphocytic pleocytosis and/or elevated protein level. Meningeal biopsies have been obtained in some cases, with results ranging from normal or benign thickening to marked thickening with reactive arachnoidal and fibrotic changes.

In this case report, we describe a patient whose chief clinical manifestation was a diffuse severe encephalopathy with marked depression of consciousness. Until now, the syndrome of intracranial hypotension has not been
Intracranial hypotension with severe encephalopathy

FIG. 1. Midsagittal T₁-weighted MR images. Left: Image obtained in the patient before placement of the blood patch and fluid resuscitation demonstrating descent of the thalamus into the posterior fossa (small arrowhead), compression of the pons against the clivus (white arrow), and tonsillar herniation (large black arrowhead). Right: Following treatment there is reelevation of the midline structures with return of the prepontine cistern (white arrow).

Case Report

History. This 40-year-old woman presented with a progressive cognitive decline of 2 to 4 weeks' duration. She was well until approximately 1 month before admission when she developed new diffuse headaches that were typically worse when she was in an upright or sitting position and were relieved when she was supine. This continued until approximately 2 weeks before admission when she suffered a brief generalized seizure. She was admitted to another hospital where she underwent computerized tomography (CT) scanning of the head and electroencephalographic (EEG) monitoring. Results of the CT scanning and EEG studies were normal. She was placed on a course of phenytoin and discharged. The patient continued to complain of severe headache, lethargy, nausea, and vomiting. She was seen by a neurosurgeon who obtained an MR image showing diffuse pachymeningitis (Fig. 2), left sphenoid sinusitis, and right maxillary sinusitis; the latter prompted the patient's referral to the otorhinolaryngology department. When the symptoms persisted the patient underwent lumbar puncture; the results were an opening pressure of 9.5 cm H₂O, a white blood cell count of 2 × 10⁶/L, and normal protein and glucose levels. Subsequently, the patient's condition deteriorated markedly. She progressed from confusion to obtundation with minimal verbal output and a marked gait ataxia, which rendered her unable to walk.

On presentation to our hospital the patient was found to have a history of rheumatoid arthritis that had not required treatment for longer than 20 years. There was no history of known malignancy, fevers, CSF rhinorrhea or otorrhea, recent flare of arthritis or back pain, further seizures, or obvious focal neurological deficit. The patient had fallen without obvious head trauma.

Examination. On examination, the patient was clearly obtunded with her eyes open only on vigorous voice stimulation accompanied by gentle shaking. Verbal output was slow, dysarthric, and limited to one- or two-word responses. Simple commands were obeyed at times with repetitive stimulation. Cranial nerve and motor examinations were unremarkable. Reflexes were mildly brisk. The Babinski's response was bilaterally extensor.

Initial evaluation included: 1) repeated lumbar puncture, which revealed very low pressure, a white blood cell count of 3 × 10⁶/L, a red blood cell count of 3 × 10¹²/L, a protein level of 39 mg/dl, and a glucose level of 55 mg/dl; 2) EEG monitoring, which displayed diffuse slowing with rare left temporal sharp waves; 3) serological...
examination, which showed a moderately elevated erythrocyte sedimentation rate of 40 mm/hour and an antinuclear antibody/titer ratio of more than 1:160; and 4) repeated MR imaging of the brain with and without addition of a contrast agent. The MR images were read (misread) as showing diffuse cerebral edema with impending herniation.

Working Diagnosis and Initial Treatment. A working diagnosis of cerebral vasculitis was established, and the patient began treatment that included high-dose corticosteroids and cyclophosphamide (Cytoxan). She was also treated for apparent diffuse cerebral edema with fluid restriction, osmotic diuresis, and loop diuresis.

The results of an open meningeal/brain biopsy were normal except for thickening of the meninges and evidence of subacute subdural hematoma. The subdural hematoma was not under pressure at surgery.

Reevaluation, Diagnosis, and Treatment. When the patient did not respond to induced diuresis and aggressive antiinflammatory treatment, an intracranial pressure monitoring device (Camino; NeuroCare, Inc., San Diego, CA) was placed. When consistently low intracranial pressures were measured (4–9 mm Hg), the patient’s clinical presentation and imaging studies were reevaluated. A myelogram was obtained that failed to demonstrate a site of the CSF leakage. Nevertheless, treatment for intracranial hypotension was instituted including recumbency (mild Trendelenburg position), intravenous hydration, and lumbar epidural blood patch placement. Placement of the blood patch resulted in an immediate rise in intracranial pressure to normal values. During the ensuing 12 to 24 hours, the patient improved markedly. Her clinical improvement correlated well with her intracranial pressures, which rose to normal values.

Posttreatment Course. One day following placement of the blood patch, the patient regained a normal mental status. A post-“CSF resuscitation” MR image demonstrated reelevation of midline cerebral structures to their normal position (Fig. 1 right). Three weeks after admission, the patient was discharged home without neurological deficit and feeling well.

Discussion

The average human brain weighs approximately 1400 g in air. It has been estimated that when floating freely in CSF, the human brain has an effective weight (that is, specific gravity) of approximately 50 g. Given the paucity of true connective tissue elements within the brain parenchyma, the brain depends greatly on the antigravity effect of the CSF to maintain its delicate structure. Robbed of this buoyancy, the brain may sag, collapsing in on itself. Often the supratentorial vacuum is then filled by expansion of the subdural space with hygroma fluid and/or blood. The protean neurological consequences of this structural shift are well illustrated by this and other cases of intracranial hypotension that have been documented in the literature.

The form of intracranial hypotension described here is probably pathophysiologically distinct from the over-shunting that is associated with postural headache and subdural hematoma. More severe neurological signs and symptoms are exceedingly rare in this relatively common syndrome. This difference in symptoms may be attributable to the relative pressure differential between the supratentorial and infratentorial compartments: in overshunting, the supratentorial pressure is low, whereas in the syndrome of intracranial hypotension, the infratentorial pressure is low. The latter promotes the dramatic descent of midline structures seen in Fig. 1 left.

The syndrome of intracranial hypotension remains an uncommon and probably underdiagnosed condition. It is, nonetheless, critical to make the diagnosis early because treatment is strikingly opposite to that of alternative diagnoses. It should be emphasized that CT and MR images in cases of intracranial hypotension may be misread to show diffuse cerebral edema with transtentorial herniation because of the compression of sulci and basal cisterns (Fig. 3). In effect, herniation or brain shift is part of the problem in intracranial hypotension because of infratentorial hypotension, rather than supratentorial hypertension. The syndrome may also be confused with Chiari type I malformation because of the prominent tonsillar herniation in both syndromes. In our view, the diagnosis should be suspected and treated empirically when the three cardinal features of the syndrome are apparent: 1) postural headache; 2) pachymeningitis; and 3) descent of midline structures on midsagittal MR images. The absence of intraparenchymal abnormalities such as white matter changes on T2-weighted MR images is an important negative finding, whereas the presence of bilateral subdural hygromas will frequently support the diagnosis.

Treatment is aimed at restoring CSF volume, rather than reducing brain edema and inflammation. Consequently, patients should be placed on strict flat bed rest (even mild Trendelenburg’s position) and receive copious intravenous hydration. In some cases continuous epidural infusions of saline has been effective. If identified, the specific source of CSF leakage should be treated. In the case described in this report, placement of a lumbar epidural blood patch was sufficient, although in the case of a Tarlov’s cyst or dural laceration, neurosurgical repair may be necessary. Medical therapy consisting of cortico-
steroids or caffeine administration has been useful in some cases.\(^3\) If subdural hygromas or hematomas are present, they will usually resolve spontaneously after normal CSF volume has been restored.\(^4\) However, if a significant amount of blood is present, the hematoma may require specific treatment itself.

The case described here adds to the previously known clinical spectrum of intracranial hypotension by demonstrating that patients may present with severe diffuse encephalopathy producing obtundation. In the present case the initial source of CSF leak was never identified. Radionuclide CSF imaging may have been a better test than the myelography that was performed. Clearly, the repeated punctures of the lumbar cistern performed during the course of the patient’s illness served only to exacerbate her condition. In view of the patient’s elevated erythrocyte sedimentation rate and antinuclear antibody ratio, a rheumatological flair may have been the trigger for this patient’s seizure and headache. This may have caused an acute rise in intracranial pressure and an occult dural tear. Alternatively, the intracranial hypotension in this case may have been totally iatrogenic, related to successive violations of the lumbar cistern.

Regardless of the initial cause, once established the intracranial hypotension became the predominant illness that required intensive care observation and responded rapidly to specific treatment both clinically and radio graphically.

References


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Address reprint requests to: Carter E. Beck, M.D., Department of Neurosurgery, MC: 5327, Stanford University Medical Center, 300 Pasteur Drive, Stanford, California 94305.