Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension

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MOST spinal cerebrospinal fluid (CSF) leaks are either iatrogenic, occurring after lumbar puncture or spinal surgery, or they may be due to a dural laceration caused by a blunt or penetrating spinal trauma. Spontaneous spinal CSF leaks are much less common but are now increasingly recognized as a cause of postural headaches associated with intracranial hypotension.1

In this report, we review our experience with 11 patients who had spontaneous intracranial hypotension caused by radiographically confirmed spinal CSF leaks. Four of the patients were treated surgically.

Clinical Material and Methods

The study population consisted of a consecutive group of patients who had a spontaneous CSF leak that was diagnosed by computerized tomographic (CT) myelography or radionucleotide cisternography at our institution between 1986 and 1995. A review was made of all of these patients’ medical records and imaging studies.

During the 10-year period under study, 11 patients with a spontaneous spinal CSF leak were identified. The mean age of these six women and five men was 38 years (range 22–51 years) at the time of diagnosis. Clinical and radiographic characteristics of the study population are summarized in Tables 1 and 2.

Results

Clinical Manifestations

A postural headache was the most characteristic and debilitating symptom in all patients and was often associated with neck stiffness and tenderness. These headaches had been present over a period of between 1 week and 9 years prior to the diagnosis of intracranial hypotension (median 1 month). Only one patient presented with...
headache and neck tenderness alone and 10 had additional symptoms at some time during their clinical course, including nausea and vomiting in five patients, sixth cranial-nerve paresis in four, local back pain in four, tinnitus in two, and seventh cranial nerve paresis, fifth cranial-nerve dysfunction, phonophobia, visual blurring, and bowel and bladder incontinence in one patient each. Headache was the initial symptom in seven patients, whereas local back pain of acute onset, corresponding to the approximate level of the CSF leak, preceded the onset of headache in four patients.

Cerebrospinal Fluid Examination

Most patients had more than two lumbar punctures performed with variable readings of CSF pressure over time, but nine patients had at least one opening pressure of 6 cm H$_2$O or less (including “dry taps” in five patients), another had an opening pressure of 9 cm H$_2$O, and one patient had an opening pressure of 10 cm H$_2$O. Similarly, variable values for cell count and protein content were obtained over time. Examination of the CSF revealed a normal cell count and total protein content in only one patient, whereas on at least one occasion pleocytosis (>5 mononuclear cells/µl) was found in five patients (maximum range 8–33 cells/µl), and elevated total protein (>45 mg/dl) in 10 patients (maximum range 48–453 mg/dl).

Imaging Studies

Spinal Imaging. Indium-111 radionucleotide cisternography was performed in 10 patients, CT myelography in nine patients, and magnetic resonance (MR) imaging in six patients (Figs. 1–3).

The location of the CSF leak was in the cervical spine in two patients, the cervicothoracic junction in three patients, the thoracic spine in five patients, and the lumbar spine in one patient. The radiographic studies indicated the presence of a meningeal diverticulum as a cause of the leak in four patients and this was confirmed at surgery in three patients. However, CT myelography failed to dem-
onstrate two meningeal diverticula found at surgical exploration of a thoracic CSF leak in another patient. In most patients, the CSF leak was rather diffuse and the exact location of the leak could not be determined. In one patient, CT myelography demonstrated extension of contrast material into the paraspinal soft tissues (Fig. 3).

Radionucleotide cisternography invariably showed early accumulation of nucleotide in the bladder, slow ascent along the spinal axis, and less than expected activity over the cerebral convexities in all patients, consistent with the diagnosis of intracranial hypotension. However, among the 10 patients who underwent radionucleotide cisternography, this study failed to identify CSF leaks diagnosed by CT myelography in three patients, for a false negative rate of 30%.

Four patients initially underwent MR imaging studies that were suggestive of a spinal CSF leak and the location of the leak was confirmed by radionucleotide cisternography or CT myelography. However, in two other patients with a documented CSF leak, MR imaging of the spine was normal.

There was no discrepancy in the location of the spinal CSF leak among different imaging studies that were able to demonstrate a leak.

### Intracranial Imaging

Craniocerebral MR imaging with gadolinium enhancement was performed in eight patients, non-enhanced MR imaging in one patient, and CT only in two patients.

Enhancement of the meninges after contrast administration, the most characteristic radiographic finding in intracranial hypotension, was found in all eight patients who were examined with gadolinium-enhanced MR imaging. Subdural fluid collections, consistent with hygroma, were found in five patients. None of these abnormal fluid collections was thicker than 1 cm and none had an associated mass effect, except for a subdural fluid collection that developed after a trephine craniotomy for meningeal biopsy performed elsewhere. This subdural hematoma was unilateral and contiguous with the biopsy site. All subdural fluid collections were managed nonsurgically.

Caudal displacement of the cerebellum, resembling a Chiari I malformation, usually associated with other signs of “brain sagging,” was present in five of nine patients examined with MR imaging. The cerebellar tonsils descended to just a few millimeters below the foramen magnum in three patients, to the level of the arch of C-1 in one patient, and down to C-2 in one patient. This last patient had undergone previous decompression of a presumed

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Cranial Imaging</th>
<th>MR Imaging</th>
<th>Radionucleotide Cisternography</th>
<th>CT Myelography</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>meningeal enhancement</td>
<td>not done</td>
<td>no leak identified</td>
<td>extrathecal contrast ventral to cord from C2–T10, maximal at C6–7</td>
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<tr>
<td>2</td>
<td>bilat subdural fluid collections</td>
<td>normal</td>
<td>increased activity of tracer at approximately T-5</td>
<td>not done</td>
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<tr>
<td>3</td>
<td>meningeal enhancement; downward cerebellar displacement (C-2)</td>
<td>not done</td>
<td>increased activity of tracer at upper lumbar region</td>
<td>abnormal contrast at L-2 neural foramens indicating meningeal diverticulum</td>
</tr>
<tr>
<td>4</td>
<td>meningeal enhancement; bilat subdural fluid collections</td>
<td>extrathecal CSF signal along entire thoracic spine</td>
<td>not done</td>
<td>circumferential extrathecal contrast from C4–L2, maximal at approximately T-8</td>
</tr>
<tr>
<td>5</td>
<td>meningeal enhancement; downward cerebellar displacement (2 mm)</td>
<td>normal</td>
<td>increased activity of tracer at cervicothoracic junction</td>
<td>circumferential extrathecal contrast from C2–T6, maximal at cervicothoracic junction</td>
</tr>
<tr>
<td>6</td>
<td>meningeal enhancement; unilat subdural fluid collection; downward cerebellar displacement (2 mm)</td>
<td>extrathecal CSF signal at upper thoracic spine</td>
<td>no leak identified</td>
<td>extrathecal contrast ventral and lateral to cord from T1–L2, maximal between T-4 &amp; T-8, indicating complex meningeal diverticulum</td>
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<tr>
<td>7</td>
<td>meningeal enhancement; downward cerebellar displacement (C-1)</td>
<td>not done</td>
<td>increased activity of tracer at lower thoracic region</td>
<td>extrathecal contrast unilateral to cord from T10–12</td>
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<td>8</td>
<td>meningeal enhancement; bilat subdural fluid collections</td>
<td>abnormal CSF signal in lt T-1 neural foramen indicating meningeal diverticulum</td>
<td>increased activity of tracer at cervicothoracic junction</td>
<td>not done</td>
</tr>
<tr>
<td>9</td>
<td>normal CT scan</td>
<td>not done</td>
<td>increased activity of tracer at lower thoracic region</td>
<td>abnormal contrast at T-11 neural foramen indicating meningeal diverticulum</td>
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<td>10</td>
<td>meningeal enhancement; bilat subdural fluid collections; downward cerebellar displacement (3 mm)</td>
<td>not done</td>
<td>no leak identified</td>
<td>extrathecal contrast ventral to cord from C2–4</td>
</tr>
<tr>
<td>11</td>
<td>normal CT scan</td>
<td>extrathecal CSF signal along cervical and upper thoracic spine</td>
<td>increased activity of tracer at cervicothoracic junction</td>
<td>extrathecal contrast ventral to cord from C2–T1 &amp; dorsal to cord from T1–4</td>
</tr>
</tbody>
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* Abreviations: CSF = cerebrospinal fluid; CT = computerized tomography; MR = magnetic resonance.
† Only an unenhanced cranial MR imaging study was performed.
Chiari I malformation and developed marked distortion of the brainstem and craniomedullary junction.

**Associated Findings of an Underlying Connective Tissue Disorder**

In four patients, an underlying connective tissue disease was strongly suspected, although no definite diagnosis could be made. Two patients possessed an unusually tall and lanky (“marfanoid”) build and had moderate joint hypermobility of the digits. One of these patients also had a high arched palate and unusually thin skin. Two other patients had suffered a spontaneous detachment of the retina at an early age (17 and 28 years, respectively).

**Treatment and Outcome**

One patient received supportive measures only (bed rest and a generous oral fluid intake) with resolution of symptoms over a period of several weeks. Two patients received temporary benefit from a short course of oral corticosteroids.

Epidural saline infusions were used in three patients, with sustained immediate relief of symptoms in one patient and minimal relief in the two other patients, who gradually improved over a subsequent period of 1 month and 2 years, respectively.

A lumbar epidural blood patch was given to three patients with CSF leaks in the cervical or thoracic spine; complete and permanent relief of symptoms was obtained in one patient and temporary benefit in the other two patients. One of those two patients subsequently underwent treatment with a cervicothoracic blood patch and achieved complete and sustained relief of symptoms. Two subsequent thoracic blood patches were administered to the other patient with transient effect only and she underwent surgical exploration of her CSF leak.

Leaking meningeal diverticula were found to be the cause of the CSF leak in the four patients who underwent surgery. One patient was found to have two meningeal diverticula, although preoperative imaging had shown only a unilateral CSF leak over several spinal levels without evidence of an underlying structural anomaly. All three patients who underwent surgical ligation of a leaking meningeal diverticulum had complete resolution of symptoms within a few days postoperatively. At the time of surgery, one patient was found to have an extensive complex of meningeal diverticula extending over the entire thoracic spine that could not be ligated or repaired. However, this patient has been free of headache during 2 months of follow up, possibly as a result of surrounding the diverticula with blood-soaked Gelfoam.

Follow-up cranial MR imaging was performed in six patients, showing resorption of subdural fluid collections, improvement or resolution of meningeal enhancement, and improvement or resolution of downward displacement of the brain. All of these patients also improved clinically at the time of the follow-up study. The shortest interval between treatment of the CSF leak and improvement of meningeal enhancement was 4 days.

**Fig. 1.** Indium-111 radionucleotide cisternograms obtained 8 hours after injection showing abnormal uptake of tracer at the cervicothoracic junction (left; arrow) in Case 5 and along a lower thoracic nerve root (right; arrow) in Case 7, suggesting a cerebrospinal fluid leak.

**Fig. 2.** Axial (left) and coronal (right) T2-weighted magnetic resonance images displaying an abnormal collection of cerebrospinal fluid (CSF) in the left T-1 nerve-root foramen (arrows) in Case 8, representing a focal CSF leak or meningeal diverticulum.

**Fig. 3.** Postmyelography computerized tomography scans showing extrathecal contrast extending bilaterally along the C-7 nerve roots into the paraspinous soft tissues in Case 5 (left) and sharply demarcated extrathecal collections of contrast along the ventral and lateral surfaces of the spinal canal at the T6–7 level in Case 6 (right).
Illustrative Case Report

Case 3. This man initially presented at age 33 years with a 1-year history of progressive, daily occipital headaches. These headaches would invariably go away if he would lie down. A gastroenterological evaluation for associated nausea and vomiting was normal.

Examination. General medical and neurological examinations were normal. Magnetic resonance studies of the head without gadolinium administration revealed that the cerebellar tonsils extended to the level of C-1, but the position of the fourth ventricle was normal. Several lumbar punctures and CSF studies were performed with opening pressures between 0 and 14 cm H₂O, cell counts between 9 and 21/µl, and total protein between 48 and 54 mg/dl. The patient was treated with corticosteroids but showed no improvement. Because of the patient’s persistent headaches a decompression of the presumed Chiari I malformation was performed.

The patient recovered well from the surgery and experienced a gradual improvement of the headaches for approximately 4 years; although they never resolved, he was able to lead a relatively normal lifestyle. However, the positional headaches worsened again over the ensuing 3 years. The headaches had been associated with nausea and vomiting, but now the patient also developed hiccups, dizziness, and bowel and bladder incontinence. Neurological examination showed generalized hyperreflexia. Magnetic resonance imaging revealed diffuse meningeal enhancement, displacement of the cerebellar tonsils to the level of C-2, distortion of the craniomedullary junction, and generalized “sagging” of the brain (Fig. 4). A lumbar puncture made with the patient in the lateral decubitus position showed an opening pressure of 6 cm H₂O, total protein of 60 mg/dl, and four nucleated cells/µl. Indium-111 radionucleotide cisternography displayed minimal migration of tracer over the cerebral convexities and an abnormal uptake of tracer in the upper lumbar spine region on the left (Fig. 5). Computerized tomographic myelography was suggestive of a meningeal diverticulum in the left L-2 neural foramen with remodeling of the L-2 pedicle, indicating a long-standing process (Fig. 6 upper). In retrospect, an abdominal CT scan performed 8 years previously for the evaluation of associated nausea and vomiting showed the presence of this diverticulum (Fig. 6 lower).

Operation. A left L-2 hemilaminectomy and total facetectomy were performed; extradural CSF was encountered immediately after removal of the ligamentum flavum. A meningeal diverticulum with multiple outpouchings was found enveloping the L-2 nerve root. There was significant bleeding from Batson’s plexus around the diverticulum, which had eroded the L-2 pedicle. The diverticulum was ligated circumferentially with several sutures without compromising the nerve root. There was no evidence of ongoing CSF leakage.

Postoperative Course. The patient recovered well from surgery with complete resolution of all his symptoms. Leg strength and sensation remained normal. One week after surgery, he noted occasional mild visual blurring. Six weeks postoperatively, examination showed bilateral papilledema with a hemorrhage inferior to the optic disc on the left. The visual symptoms resolved spontaneously and 4 months later the papilledema improved considerably and the hemorrhage resolved. Magnetic resonance imaging of the head revealed resolution of the meningeal enhancement, elevation of the cerebellar tonsils, and marked improvement of the cerebral displacement (Fig. 4).

We postulate that the development of papilledema in this patient may have been due to the sudden interruption of the abnormal pathway of CSF resorption, which had been present for many years.

Discussion

The syndrome of spontaneous intracranial hypotension is characterized by a postural headache that may be associated with a variety of symptoms including posterior neck pain or stiffness, nausea, vomiting, diplopia, visual blurring, tinnitus, vertigo, and local back pain. The diagnosis is confirmed by lumbar puncture, which reveals a low CSF pressure. However, variable readings of CSF pressure may be obtained over time, possibly indicating that the CSF leak is intermittent. Examination of the CSF itself often shows mild elevations of total protein and increased cell count.

Characteristically, cranial MR imaging studies in pa-
tients with intracranial hypotension reveal diffuse enhancement of the meninges and meningeal biopsies usually show a thin layer of fibroblasts in the subdural zone with small thin-walled blood vessels in an amorphous matrix and no evidence of inflammation or any abnormal cells. The most important intracranial complications of intracranial hypotension are subdural hematoma or hygroma and caudal displacement of the cerebellar tonsils. These complications are not believed to be the cause of the orthostatic headache, but rather secondary to the loss of buoyancy and resultant downward displacement and settling of the brain on the cranial floor. Caudal displacement of the cerebellar tonsils generally is mild and rarely extends more than several millimeters below the level of the foramen magnum. The cerebellar herniation seen in one of our patients down to the C-2 level with marked distortion of the brainstem and craniomedullary junction is distinctly uncommon and may have been due to the unusual chronicity of the spinal CSF leak (9 years). In patients with intracranial hypotension, sixth cranial-nerve paresis as well as the fifth and seventh cranial-nerve dysfunction observed in one of our patients may be due to traction on the nerve secondary to the “sagging” of the brain.

Etiology and Radiographic Features of Spontaneous Intracranial Hypotension

Nucleotide cisternography has been shown to be fairly characteristic in patients with spontaneous intracranial hypotension, showing early accumulation of nucleotide in the bladder and less activity than expected over the cerebral convexities, which are suggestive of unusually rapid uptake of the tracer in the bloodstream. Occasionally, a spinal CSF leak is demonstrated. The study should be performed with nasal pledgets in place to help exclude a spontaneous cranial CSF leak. Spontaneous intracranial hypotension may also result from a CSF leak through the skull base but this is rare if the patient has no history of rhinorrhea or otorrhea. If the nucleotide study does not show any leak but is suggestive of rapid uptake of tracer, this does not necessarily indicate the presence of generalized CSF hyperabsorption but a more likely alternative explanation is the presence of a leak below the limit of resolution of the study.

Myelography with subsequent CT scanning through the area suspected of harboring a CSF leak is required to localize the leak more precisely and may reveal an underlying anatomical defect causing the leak such as a meningeal diverticulum. If myelography is performed without the benefit of a previous imaging study that demonstrates

Fig. 5. Indium-111 radionucleotide cisternograms obtained 12 hours after injection showing abnormal activity of tracer in the upper lumbar spine on the left (arrow). Also note poor migration of tracer over the cerebral convexities.

Fig. 6. Upper Panels: Postmyelography computerized tomography (CT) scans showing a meningeal diverticulum in the left L-2 neural foramen with remodeling of the L-2 pedicle. Lower Panel: An abdominal CT scan performed 8 years previously shows the same lesion (arrow).
the approximate location of the leak, then CT slices should be obtained through the skull base and along the entire spinal axis at each spinal level. In our study, however, concordance for the presence of a spinal CSF leak between nucleotide cisternography and CT myelography was poor and the false negative rate of nucleotide cisternography was high (30%).

Occasionally, MR imaging studies of the spine may demonstrate extravethal CSF collections, which suggest the presence of a CSF leak. In our study, the false negative rate for MR imaging also appeared to be significant (33%), but our experience with spinal MR imaging in patients with spinal CSF leaks is limited.

Although leaking meningeal cysts or simple dural tears frequently have been mentioned as the likely cause of spontaneous intracranial hypotension, radiographic documentation of a spinal CSF leak has been uncommon and surgical confirmation of the CSF leak has been reported only rarely. In our four patients who underwent surgical treatment for their intracranial hypotension, leaking meningeal diverticula were found in all patients.

**Spinal Meningeal Diverticula**

Spinal meningeal diverticula are abnormal outpouchings of the common dural sac, the spinal arachnoid, or the nerve root sheath. These diverticula have been reported to be the cause of low-back pain, radiculopathy, and myelopathy and may be associated with spontaneous spinal cord herniation. Recently, Nabors, et al. reviewed their experience with meningeal diverticula and classified them into three distinct types: extradural diverticula without spinal nerve root fibers (Type I); extradural diverticula with spinal nerve root fibers (Type II, Tarlov’s cyst); and intradural cysts (Type III). Of our four patients with surgically confirmed meningeal diverticula, two patients had a Type II lesion; one patient had two Type I lesions, both arising from the axilla of the nerve root; and one patient had an extensive complex of meningeal diverticula, which was extradural and may have involved spinal nerve root fibers.

The etiology of most meningeal diverticula is unknown, and it is not clear whether these are congenital or acquired lesions. It is likely, however, that, at least in some cases, an underlying weakness of the spinal meninges is involved, predisposing the patient to develop meningeal diverticula. Several heritable connective tissue disorders have been associated with spinal meningeal diverticula, such as Marfan syndrome and neurofibromatosis. Two of our patients with surgically confirmed meningeal diverticula had an unusual “marfanoid” body habitus and joint hypermobility, suggesting the presence of a generalized connective tissue disorder, but the patients’ phenotype did not fulfill the criteria for any of the known connective tissue disorders. In addition, two other patients had suffered a retinal detachment at an early age without a prior history of trauma. Spontaneous retinal detachment in the young has been reported in heritable connective tissue disorders such as Stickler, Marfan, and Ehlers–Danlos syndromes, and osteogenesis imperfecta; however, our two patients had none of the other stigmata associated with these connective tissue disorders. To our knowledge, the coexistence of spontaneous spinal CSF leak and spontaneous retinal detachment has not been described previously; however, this association indicates an abnormality of a connective tissue component common to both the retina and the spinal meninges.

**Treatment Options in Spontaneous Intracranial Hypotension**

Fortunately, most cases of intracranial hypotension resolve spontaneously and a variety of treatment options are available for patients with persistent symptoms. Conservative treatment consists of bed rest and oral hydration. Medical treatment options include caffeine, mineralocorticoids, glucocorticoids, and parenteral fluid and salt infusion. A more direct and, in our experience, probably more efficacious method of treatment is the epidural blood patch, as originally described by Gormley for the treatment of postlumbar-puncture headache. However, the benefit attained with a lumbar epidural blood patch is often only temporary, particularly when performed in the lumbar spine for CSF leaks at the cervical or thoracic spinal levels. An epidural blood patch can also be performed in the cervical or thoracic spine but with increased risk; under such circumstances, consideration should be given to intrathecal or epidural saline infusions.

Surgery may be considered for young patients with persistent symptoms who have a demonstrable leak at a location that is surgically accessible with low risk. In our limited experience, the surgical treatment of CSF leaks associated with meningeal diverticula has generally been satisfactory. Most meningeal diverticula without spinal nerve root fibers can be treated with simple ligation of the neck of the diverticulum with suture, provided the size of the diverticulum is not prohibitive. The surgical treatment of meningeal diverticula containing spinal nerve root fibers is more complex when the root is believed to be eloquent, and an attempt can be made to plicate and reconstruct the diverticulum or, at least theoretically, ligate the sac around the nerve root without compromising the root.

**Conclusions**

Spontaneous spinal CSF leaks are uncommonly encountered in neurosurgical practice, but they are increasingly recognized as a cause of spontaneous intracranial hypotension. Most spinal CSF leaks are located at the cervicothoracic junction or in the thoracic spine and they may be associated with meningeal diverticula. Although radionucleotide cisternography and MR imaging play a definite role, the radiographic study of choice is CT myelography. The disease is often self-limiting, but in cases with protracted symptoms we suggest that surgical ligation of leaking meningeal diverticula can be accomplished with satisfactory results. An underlying connective tissue disorder may be present in some patients who present with a spontaneous spinal CSF leak.

**References**

3. Bourekas EC, Lewin JS, Lanzieri CF: Postcontrast meningeal...
Spontaneous spinal CSF leaks


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