OBJECTIVE AND IMPORTANCE: Spontaneous intracranial hypotension is an increasingly recognized cause of postural headache. However, appropriate management of obtundation caused by intracranial hypotension is not well defined.

CLINICAL PRESENTATION: A 43-year-old man presented with postural headache followed by rapid decline in mental status. Imaging findings were consistent with the diagnosis of spontaneous intracranial hypotension, with bilateral subdural hematomas, pachymeningeal enhancement, and caudal displacement of posterior fossa structures and optic chiasm.

INTERVENTION: Despite treatment with lumbar epidural blood patch, worsening stupor necessitated intubation and mechanical ventilation. Contrast-enhanced magnetic resonance imaging and computed tomographic myelography of the spine failed to demonstrate the site of cerebrospinal fluid fistula. The enlarging subdural fluid collections were drained, and a ventriculostomy was performed. Postoperatively, the patient remained semicomatose. To restore intraspinal and intracranial pressures, intrathecal infusion of saline was initiated. After several hours of lumbar saline infusion, lumbar and intracranial pressures normalized, and the patient’s stupor resolved rapidly. Repeat computed tomographic myelography accomplished via C1–C2 puncture demonstrated a large ventrolateral T1–T3 leak, which was treated successfully with a thoracic epidural blood patch. Follow-up magnetic resonance imaging demonstrated resolution of intracranial hypotension, and the patient was discharged in excellent condition.

CONCLUSION: Spontaneous intracranial hypotension may cause a decline of mental status and require lumbar intrathecal saline infusion to arrest or reverse impending central (transtentorial) herniation. This case demonstrates the use of simultaneous monitoring of lumbar and intracranial pressures to appropriately titrate the infusion and document resolution of intracranial hypotension. Maneuvers aimed at sealing the cerebrospinal fluid fistula then can be performed in a less emergent fashion after the patient’s mental status has stabilized.

KEY WORDS: Blood patch, Cerebrospinal fluid leak, Intracranial hypotension, Intrathecal, Lumbar infusion, Obtundation

We discuss the appropriate diagnosis and management of SIH and demonstrate from this case report that such symptoms of impending transtentorial herniation can be reversible.

CASE REPORT

A 43-year-old man with a medical history only of sinusitis presented with subacute onset of headache (3 wk), which was postural initially (worse at upright posture and improved...
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when lying down) and then became constant. This was followed by the development of horizontal diplopia and a progressive, rapid decline in gait and mental status that prompted hospital admission. At admission, he was stuporous with limited neck flexion, increased lower extremity tone, and extensor plantar responses. A brain computed tomographic (CT) scan obtained at another institution demonstrated evolving bilateral subdural hygromas and descent of the brain into the posterior fossa with effacement of the basal cisterns (Fig. 1A). MRI of the brain performed at admission demonstrated large bilateral subdural hygromas, marked descent of the brain with effacement of the basal cisterns by descent of the midbrain and brainstem onto the clivus, cerebellar tonsillar herniation into the foramen magnum, descent of the optic chiasm onto the dorsum sellae, and diffuse dural contrast enhancement (Fig. 1B and C). MRI of the entire spine was normal, demonstrating no evidence of an obvious perineural cyst or cerebrospinal fluid (CSF) leak. Magnetic resonance venography revealed a dominant right transverse sinus, and diffusion-weighted imaging revealed no abnormalities.

In view of the patient’s progressive obtundation and the MRI findings suggesting intracranial hypotension, emergent treatment with a large-volume (30-ml) lumbar (L3–L4) epidural blood patch was performed, and the patient then was moved into a reverse Trendelenburg position for 2 hours. After the procedure, he had transient improvement in mental status, became oriented to person and place, and followed commands; however, his stupor returned within 12 hours. A second 30-ml lumbar blood patch was placed, which also resulted in transient improvement. However, the patient developed hiccups and labored breathing and became unresponsive. His pupils were unreactive bilaterally, and he had primitive grasp reflexes bilaterally. He could no longer protect his airway, and abruptly he required intubation and mechanical ventilation.

Immediate consideration was given to intrathecal saline infusion; therefore, a lumbar subarachnoid drain was placed under fluoroscopic guidance. At the same time, an emergent CT myelogram was performed in an attempt to localize the presumed spontaneous CSF leak. This demonstrated sequelae of previous lumbar epidural blood patches and abnormal extrathecal contrast accumulation extending from C7 to T6 suggestive of a CSF fistula, but with no obvious source of leak. A follow-up brain CT scan demonstrated interval increase in the large bilateral subdural hematomas and continued signs of central herniation.

The patient was taken to surgery on an emergent basis to drain the large bilateral subdural fluid collections. Burr holes were made bilaterally 12 cm posterior to the nasion and 3 cm lateral to the midline. Thick dural membranes were encountered and coagulated with bipolar cautery. The dura was opened in cruciate fashion, and a moderate amount (approximately 50 ml) of xanthochromic fluid was expressed at very low pressure (white blood cells, 733/μl; red blood cells, 820,000/μl; and glucose, 43 mg/dl). This area was irrigated copiously until clear. Lowering the head of the bed clearly increased the egress of fluid. Notably, the brain did not appear to rise or change position after drainage of these collections. A right frontal external ventricular drain catheter was placed, and CSF returned to a very low pressure (0 cm H2O).

Postoperatively, the patient was taken to the neurosurgical intensive care unit. Despite drainage of the subdural fluid, he remained severely obtunded with no improvement in mental status. Both the lumbar subarachnoid catheter and the external ventricular drain were connected to pressure transducers. In an attempt to restore intraspinal pressure and intracranial pressure (ICP), intrathecal infusion of preservative-free saline was initiated via lumbar catheter, with a bolus of 50 ml and then continuous infusion at a rate of 30 ml/h using a Medfusion pump (Medfusion Systems, Norcross, GA). ICP was monitored continuously, and subarachnoid lumbar pressure (LP) was monitored intermittently.

Initial LP and ICP were near zero with no waveform (Table 1). After 3 hours of lumbar saline infusion at 30 ml/h, LP had gradually increased to 15 mm H2O, but ICP remained at 0. After 2 hours with LP in the range of 15 to 17 cm H2O, there was an abrupt increase in ICP to 18 cm H2O and resumption of a normal ICP waveform (Table 1). This was accompanied by an immediate and dramatic improvement in mental status. The patient began to move around purposefully, and his pupils became reactive bilaterally. Thereafter, both LP and ICP correlated within 3 mm H2O, and normal waveforms varied with the cardiac cycle. As LP and ICP increased above 20 mm

![FIGURE 1. CT scan (A) obtained 2 days before admission and MRI scans (B and C) obtained at admission. The patient was obtunded at this time. A, axial noncontrast CT scan demonstrating bilateral frontal subdural fluid collections, which are similar in density to CSF and result in mild mass effect. B, sagittal precontrast T1-weighted MRI scan demonstrating downward displacement of the midbrain,pons, and optic chiasm with narrowing ofthe distance between the mamillary bodies and upperpons (arrow). Effacement of the suprasellar cistern is observed. C, coronal postcontrast T1-weighted MRI scan demonstrating diffuse dural enhancement (arrows) and right subdural effusion (arrowheads).](image-url)
H₂O, the lumbar saline infusion was stopped to avoid intracranial hypertension. Thereafter, LP and ICP slowly decreased. By 2 hours after resumption of normal ICP, the patient was briskly following commands with all four extremities. Two hours later, he was extubated, awake, alert, and fully oriented. Apart from a 3-day period of antegrade amnesia for the event and resolving horizontal diplopia and hyperreflexia, he became neurologically normal.

To better localize the CSF fistula, a repeat cervical CT myelogram was performed later that day via C₁–C₂ puncture under direct CT guidance. The patient was positioned prone on the CT scanner, and a C₁–C₂ puncture was performed using C-arm fluoroscopy. Multiple scans were performed through the cervical and thoracic levels to document more accurately the exact position of the CSF fistula. On the second set of CT scans, an accumulation of extradural contrast medium was identified to the left of midline from the ventrolateral thecal sac (Fig. 2A). The contrast medium subsequently spread cranially and caudally from beyond the T₁–T₃ levels, but no specific site of leakage was identified. Successful treatment consisted of a large-volume (30-ml) epidural blood patch placed directly at the T₄ level under CT guidance. The patient was maintained at bed rest for several days, during which time LP and ICP remained normal (ICP range, 4–21 cm H₂O).

The lumbar and intracranial catheters were removed on hospital Day 9, and the patient was discharged home in excellent condition on hospital Day 11. On the day of discharge, repeat brain MRI demonstrated decreased size of bilateral subdural hygromas, decreased dural enhancement, and elevation of the brainstem and cerebellum with restoration of basal cisterns (Fig. 2, B and C).

**DISCUSSION**

**History**

Since Quincke (44) introduced lumbar puncture in 1891, the syndrome of spinal or postlumbar puncture headache has been well described (62). The German neurologist Schaltenbrand (49, 50) was the first to describe a condition of spontaneously low or even negative CSF pressure with orthostatic headache, among other symptoms, which he termed primary SIH or “essential aliquorrhea.” A similar condition was described by Woltman (66). Subsequent investigators fur-
ther defined this syndrome of intracranial hypotension or "hypoliquorrhea" (3, 24, 26).

Etiology

It was originally theorized that the "hypo-" or "aliquorrhea" resulted from decreased CSF production or increased CSF reabsorption (49, 50). In 1976, Labadie et al. (23) used isotope cisternography to demonstrate normal CSF flow but rapid appearance of isotope in the urinary bladder, inconsistent with decreased production but consistent with either hyperabsorption or cryptic CSF leakage. Even into the early 1990s, some investigators thought hyperabsorption of CSF was responsible (38, 64), despite the prior report of a patient with a localized T5 leak that closed after epidural saline infusion (18). In 1992, Rando and Fishman (47) reported two cases of intracranial hypotension in which radioisotope cisternography demonstrated localized CSF leaks, and they hypothesized that the mechanism of CSF leak was caused by spontaneous rupture of a spinal arachnoid (Talarov's) cyst. On the basis of this report and subsequent series, it is now thought that spontaneous spinal CSF leaks are the most common cause of SIH (51, 55).

The cause of spontaneous CSF leaks is unknown but probably involves weakness of the spinal meninges (30, 39, 55, 58). Some cases of spontaneous CSF leak are associated with generalized connective tissue disorders. Meningal diverticula and SIH have been described with Marfan's syndrome (6, 11, 16, 55, 56), and meningeal diverticula also have been described in autosomal dominant polycystic kidney disease (53), neurofibromatosis Type 1 (10, 29), and Lehman syndrome (25, 42). Presence of structural dural weakness or meningeal diverticula may allow CSF to leak into the extradural space, especially after a bone spur piercing the dura (9, 63) oriatrogenic causes (e.g., thoracic or spinal surgery) (22).

Clinical Manifestations

SIH is nearly always (52) characterized by orthostatic headache that is worse with upright posture and improved during recumbency. In the presence of subdural effusions or hematoma, however, the headache may be constant and may lack postural features. In general, the onset of the headache is gradual or subacute, but some patients may present with thunderclap headache mimicking aneurysmal subarachnoid hemorrhage (59). The headache may be holocranial or localized to the frontal or occipital regions. It is probably caused by CSF volume loss and subsequent traction on the pain-sensitive intracranial dura caused by downward brain displacement. Asssociated symptoms vary and may include neck pain or stiffness, nausea, vomiting, horizontal diplopia, dizziness, hearing and visual changes, phonophobia, and photophobia (3, 5, 12, 20, 37, 51, 55, 65). Horizontal diplopia, hearing changes, and vertigo may be attributable to traction of the abducens, cochlear, and vestibular nerves. Facial numbness or pain (37, 55) may be caused by traction of the trigeminal nerve, transient visual obscurations (20) to traction of the optic apparatus, and galactorrhea (67) to traction of the pituitary stalk. Rarely, as in this patient, severe sagging of the brain may lead to signs of central (transtentorial) herniation with stupor caused by diencephalic compression (2, 43, 55).

Diagnosis

Neuroradiology is crucial in the evaluation and management of SIH, both for the initial diagnosis of SIH and for accurate localization of the site of CSF leakage. In the early 1990s, advances in MRI dramatically facilitated the diagnosis of intracranial hypotension (14, 19, 35, 40, 48). The main imaging findings of SIH on CT and MRI scans, which also were observed in our patient, are secondary to increased venous engorgement and include pachymeningeal enhancement, descent of the brain into the posterior fossa, and bilateral subdural hygromas. Other features include enlargement of the dural venous sinuses and pituitary gland and chiasmatic drooping on the dorsum sellae.

Pachymeningeal enhancement after gadolinium administration occurs in at least 90% of patients with intracranial hypotension (5, 14, 34, 35, 37, 40, 48). This is thought to result from dural venous engorgement secondary to increased intracranial blood volume after CSF volume depletion, according to the Monro-Kellie rule (14, 31). A similar mechanism involving venous hyperemia may account for enlargement of the pituitary gland (1, 32, 33). Meningeal biopsies have demonstrated thin-walled dilated blood vessels without inflammation, but this finding is unnecessary for diagnosis (13, 21, 36).

Descent of the brain into the posterior fossa, also called brain sagging or pseudo-Chiari malformation, is another characteristic of SIH. Imaging features include effacement of basal cisterns, flattening of the pons against the clivus, and hindbrain herniation with tonsillar ectopia (Fig. 1B) (14, 28, 40, 60). Tonsillar displacement into the spinal canal may prompt the diagnosis of Chiari I malformation; therefore, some patients with SIH have undergone suboccipital decompressive surgery (55).

Finally, subdural fluid collections (hematomas and hygromas) are commonly observed in patients with SIH (5, 14, 28, 40, 60). Subdural hematomas are thought to be caused by tearing of bridging veins attributable to downward brain displacement. Subdural hygromas may constitute a compensatory increase in the volume of the subdural space, again according to the Monro-Kellie rule.

CSF Findings

It is unclear whether CSF evaluation by lumbar puncture is necessary for the diagnosis of SIH. Lumbar puncture may create another leak and aggravate the syndrome (15). Typically, opening pressure of less than 60 mm H2O is documented (47), and in some cases CSF can only be obtained via Valsalva maneuver or by aspiration (3). CSF examination variably demonstrates pleocytosis, elevated protein, and xanthochromia (37, 47). In some cases, this has led to the erroneous diagnosis of aseptic meningitis or encephalitis, especially when a
Localization of the Site of CSF Fistula

Localization of the site of CSF fistula can be difficult. A typical search for the leak begins with fat-suppressed fast spin echo MRI of the spine (7), which may reveal an extradural collection of CSF or a prominent perineural cyst. Supportive evidence for a spinal CSF leak includes CSF hygroma, epidural venous engorgement and dural enhancement, and paraspinal fluid collections behind C1–C2 (7). As in our patient, however, spinal MRI often is not sensitive enough to detect the actual site of a CSF leak, although extrathecal CSF accumulations and meningeal diverticula are visible (37, 45, 47, 55). Dynamic decubitus myelography followed by CT myelography is currently the study of choice to define the exact location of a spontaneous CSF leak (55). CT myelography can demonstrate a site of extrathecal contrast accumulation or define anatomic abnormalities such as meningeal diverticula (7, 8). The majority of leaks are found at the cervicothoracic junction or in the upper thoracic spine.

Radionuclide cisternography also has been used commonly to evaluate spinal CSF leaks (6, 23, 38, 47). However, it is less sensitive than CT myelography and has poorer spatial resolution (55). It is useful in patients with cranial CSF leaks. The new modality of magnetic resonance myelography may prove useful (27, 46).

Treatment

The headache in SIH usually resolves spontaneously. Occasionally, a prolonged period of bed rest is required. Medical management of SIH is largely ineffective. With intractable persistent headache or other signs and symptoms of SIH, the treatment of choice is epidural blood patch (4, 8, 17, 37, 55). A large-volume blood patch (>20 ml) is preferable to a smaller-volume patch to cover a larger epidural territory. Because the most common site of leakage is cervicothoracic, the patient should be placed in a reverse Trendelenburg position with the head lowered approximately 30 degrees for at least 10 minutes after the large-volume patch is used. Relief of symptoms is often immediate as a result of thecal compression and increased CSF pressure. It is presumed that long-term effectiveness relates to dural tamponade or fibrosis that scars over the site of the leak. Occasionally, several large-volume blood patches are necessary for permanent benefit (8). The initial blood patch usually is placed in the lumbar spine regardless of the site of the leak because blood is known to spread over many levels (61); if ineffective, a more directed patch can be placed after more precise localization of the site of the fistula (at T4 in our patient). Other techniques that have been used include epidural saline infusion and percutaneous fibrin glue injection (18, 41).

If all other measures fail, and the leak has been well localized by imaging studies, surgery can be attempted to repair the leak. This has been especially effective in the treatment of meningeal (Tarlov’s) cysts (43, 57, 58). Ligation of the diverticulum and/or packing of the epidural space with Gelfoam (Upjohn Co., Kalamazoo, MI) or fibrin glue has been proved effective in individual cases.

Management of Obtundation

There have been two prior case reports of patients with SIH presenting with stupor. Beck et al. (2) described a 40-year-old woman with progressive cognitive decline and orthostatic headache with imaging findings of SIH. Meningeal biopsy results were normal, with evidence of subacute subdural hematoma, and an ICP monitor was placed that demonstrated low ICP. Placement of a lumbar epidural blood patch led to an immediate increase in ICP to normal values and accompanying clinical resolution. Pleasure et al. (43) described a 51-year-old man with progressive headache, somnolence, and imaging findings of SIH. He did not improve after placement of three epidural blood patches, and intrathecal saline infusion was initiated via lumbar drain at 30 ml/h. LP was monitored, and the infusion was adjusted to a target of 11 mm Hg (14.3 cm H2O). The patient rapidly improved to normal mental status with the exception of antegrade and retrograde (1 mo) memory loss. In this case, the lumbar infusion was continued at 10 to 30 ml/h during evaluation for a CSF leak. Ultimately, he had two more blood patches that did not convey lasting relief, followed by surgery, which demonstrated a T4 Tarlov's cyst that subsequently was ligated.

With these cases, our case report demonstrates that SIH may not have a benign or indolent course. In the case of rapidly declining mental status, lumbar intrathecal saline infusion may be implemented to arrest or reverse impending central (transientorial) herniation. Our case report further demonstrates that simultaneous monitoring of LP and ICP can be used to appropriately titrate the saline infusion and document resolution of intracranial hypotension. Although in some cases, lumbar saline infusion alone may be sufficient to reverse obtundation, in patients without immediate clinical response it may be valuable to monitor ICP concurrently to titrate the infusion. We also demonstrate that ICP and subarachnoid LP may not be correlated, presumably because of incisural block caused by downward displacement of cerebral structures. After several hours of lumbar saline infusion, during which time the LP slowly increased, ICP remained at 0 with no waveform, followed by an abrupt increase to 18 cm H2O with normal waveform and subsequent close correlation with the subarachnoid LP. It is presumed that this represents an “uncorking” phenomenon of the midbrain/hindbrain structures that was correlated with a dramatic improvement in the clinical condition of our patient. Although imaging studies were not immediately obtained, follow-up MRI demonstrated evidence of reversal of hindbrain herniation and restoration of the basal cisterns (Fig. 2, B and C). Thereafter, with the patient’s mental status stabilized, diagnostic and therapeutic maneuvers aimed at sealing the site of the leak were performed in a less emergent fashion and ultimately were successful at localizing and treating an upper thoracic leak.
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CONCLUSION

In summary, our case report demonstrates that spontaneous intracranial hypotension may be complicated by obtundation, that this obtundation can be reversible by lumbar intrathecal saline administration, and that simultaneous LP and ICP measurements can be useful to appropriately titrate this infusion. Appropriate localization and treatment of the spontaneous CSF leak then can be undertaken.

REFERENCES


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The authors’ patient experienced positional headaches and developed rapidly progressive rostrocaudal brain herniation with associated subdural hygromas and tonsillar foramen magnum herniation, all of which were the result of a spontaneous thoracic cerebrospinal fluid leak. Despite early suspicion of the diagnosis, the initiation of treatment with positional maneuvers, an epidural blood patch, the creation of burr holes, and ventriculostomy failed to bring about recovery. Only by infusing saline into the lumbar subarachnoid space did the condition reverse. The authors demonstrate that by transducing both intracranial and intraspinal pressure, “uncorking” of the foramen magnum temporally correlated with neurological improvement. Although a similar case from the University of California, San Francisco, was published previously (1), the current case reconfirms the lifesaving implication of timely recognition of this reversible condition.

Luckily, this clinical scenario is rare. Spontaneous cerebrospinal fluid fistulae and intracranial hypotension syndromes are well known, but such a significant, life-threatening sequela is outside most neurosurgeons’ clinical experience. Certainly, neurosurgeons are commonly asked to evaluate patients with positional and cough headaches for possible Chiari I malformations. On the basis of the case material presented here, the diagnosis of acquired rather than primary Chiari I malformation in patients who have associated subdural hygromas and in whom there is rapid neurological deterioration should be considered, because suboccipital decompression would not be the treatment of choice. It behooves neurosurgeons to recognize that such a syndrome exists and to remember that the means exist to treat it.

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BINDER ET AL.

The authors describe the cause, the pathophysiology, and the treatment of a patient with spontaneous intracranial hypotension caused profound neurological deficits. The authors diagnosed and treated this patient aggressively in a complicated case. Both the case described here and the accompanying review of the literature will help other neurosurgeons to become more aware of this diagnosis and of the benefits of lumbar intrathecal saline infusion in these cases.

Alex B. Valadka
Houston, Texas

COMMENTS

In this interesting report from the University of California, San Francisco, the authors’ patient arose phoenixlike from the ashes of deepest coma to complete recovery, which the authors attribute to a lumbar intrathecal saline infusion. Preposterous as it may seem, the case is carefully presented, and the pathophysiology is clearly explained, such that there is little doubt about the merits of the authors’ conclusions.

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tion of the causative thoracic leak, the authors’ technique proved to be quite successful, and the use of this method to correct a grave clinical condition is worthy of note. The “uncorking phenomenon” described in the Discussion connotes a rather violent movement of delicate structures and seems dangerous, but the results speak for themselves.

Warren R. Selman
Cleveland, Ohio

Spontaneous intracranial hypotension is a relatively uncommon neurosurgical entity. The case of the patient whom Binder et al. describe is an extreme example of the possible neurological sequelae of this condition. The management of this patient with a lumbar saline infusion to restore normal intracranial pressure followed by a high thoracic epidural blood patch proved to be effective and curative. This case report adds significantly to the growing literature on this rare condition. It also demonstrates that in the majority of individuals, the site of the cerebrospinal fluid fistula can be sealed with an appropriately placed epidural blood patch to obviate the need for an open surgical procedure.

Daniel F. Kelly
Los Angeles, California

Resident Traveling Fellowship in Pediatric Neurosurgery

The Joint Pediatric Neurosurgery Section of the American Association of Neurological Surgeons and Congress of Neurological Surgeons has established a traveling fellowship for residents in accredited neurosurgical training programs. The fellowship is intended to cover the traveling and living expenses for up to 1 month for a resident who wishes additional experience in pediatric neurosurgery during residency. The fellowship can be spent at any institution within North America to pursue an activity that broadens the resident’s exposure to pediatric neurosurgery, including observation at a clinical or research center, participation in a research project, or any other relevant activity. Two fellowships per year are awarded on the basis of an evaluation by a committee of the Joint Pediatric Section. The maximum fellowship stipend is $2500.

The application should include:
1. Statements defining the purpose of the proposed fellowship and an estimate of expenses that will accrue to the applicant;
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3. A letter of acceptance from the pediatric neurosurgical program where the applicant will seek the fellowship.

The deadline for application submission is October 15, 2002. The completed application should be sent to:

R. Michael Scott, M.D.
Department of Neurosurgery
The Children’s Hospital
300 Longwood Avenue, Bader 319
Boston, MA 02115

(or via email to: michael.scott@tch.harvard.edu)