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Low-Pressure Hydrocephalus and Shunt Malfunction Following a Lumbar Puncture in an Adult Reversed by an Epidural Blood Patch

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Key words
- Blood patch
- Hydrocephalus
- Low pressure
- Lumbar puncture
- Shunt malfunction

Abbreviations and Acronyms
- CSF: Cerebrospinal fluid
- CT: Computed tomography
- ICP: Intracranial pressure
- LPH: Low-pressure hydrocephalus
- VP: Ventriculoperitoneal

INTRODUCTION

Low-pressure hydrocephalus (LPH) is a rather uncommon condition in which there is ventricular dilation despite low intracranial pressures (ICPs).1,2 The symptoms perceived are identical to those usually observed in the more commonly seen high-pressure hydrocephalus, with headaches, cranial nerve dysfunction, ataxia, and disturbances of consciousness. Cerebral cerebrospinal fluid loss in the presence of altered brain viscoelastic properties has previously been suggested as the pathophysiology process leading to ventriculomegaly, despite low or negative intracranial pressures and patent shunts. More recently, cerebral venous overdrainage has been proposed as a possible explanation in the pathogenesis of LPH, although its connection to lumbar punctures in patients with shunts has not been contemplated yet. The effectiveness of epidural blood patch in the management of post-lumbar puncture LPH has been shown in children but has not been reported in adults.

CASE DESCRIPTION: Herein we detail 2 episodes of shunt malfunction in a 30-year-old female patient with a history of hydrocephalus related to a posterior fossa tumor diagnosed during childhood. In both instances, imaging studies demonstrated ventricular dilation along with perimedullary cistern enlargement and brainstem distortion, which occurred following a lumbar puncture despite a patent shunt. A lumbar blood patch was effective in both episodes, enabling resolution of the ventriculomegaly and a good outcome.

CONCLUSIONS: A blood patch can be efficient in adults with post-lumbar puncture LPH. Some symptoms may be explained by brainstem compression caused by enlarged cerebrospinal fluid spaces at the skull base. The role of cerebral venous overdrainage in the setting of post-lumbar puncture LPH is further supported.

BACKGROUND: Low-pressure hydrocephalus (LPH) is a relatively rare condition, and its presentation is similar to the classically seen high-pressure hydrocephalus, with headaches, cranial nerve dysfunction, ataxia, and disturbances of consciousness. Cerebral cerebrospinal fluid loss in the presence of altered brain viscoelastic properties has previously been suggested as the pathophysiology process leading to ventriculomegaly, despite low or negative intracranial pressures and patent shunts. More recently, cerebral venous overdrainage has been proposed as a possible explanation in the pathogenesis of LPH, although its connection to lumbar punctures in patients with shunts has not been contemplated yet. The effectiveness of epidural blood patch in the management of post-lumbar puncture LPH has been shown in children but has not been reported in adults.

CASE REPORT

A 30-year-old female patient presented to Mater Dei Hospital with a complaint of new-onset headaches. She had a medical history that included a posterior fossa medulloblastoma treated with surgery, radiotherapy, and chemotherapy when she was 10 years old, followed by a ventriculoperitoneal (VP) shunt. In the past, several shunt revisions had been performed, 2 of them related to a lumbar puncture, at different outside hospitals. The last shunt revision took place several
years ago. None of the medical records detailing these shunt revisions were available for review. Neurologic examination did not disclose any abnormalities. Imaging studies demonstrated a calcified pineal region mass, isointense on T1-weighted and fluid-attenuated inversion recovery images, mildly hyperintense on T2-weighted images, and homogeneously enhancing with gadolinium, measuring 2 cm in diameter (Figure 1A). Slight ventricles could be depicted (Figure 1A) as well as 2 ventricular catheters, with only one of them connected to a VP shunt system. Meningeal enhancement along the right cerebral convexity, suggestive of shunt overdrainage, was also observed (Figure 1A). Magnetic resonance angiogram and venography did not demonstrate major abnormalities.

A right occipital/transventricular approach was performed and a subtotal resection of the pineal mass (grade I meningioma) was achieved. An attempt to place a L4–L5 lumbar drain immediately before the craniotomy was unsuccessful as catheter advancement was not possible and approximately 10 cc of CSF was drained. She had an uneventful immediate postoperative course except for a mild left-sided homonymous hemianopsia. A routine postoperative computed tomography (CT) scan of the head demonstrated pneumocephalus but otherwise stable ventricles (Figure 1B). A few days later she complained of nausea, somnolence, and diplopia. A CT scan of the head demonstrated hydrocephalus (Figure 1C), and she was taken to the operating room for a shunt revision. Intraoperative inspection of the pre-existing medium-pressure shunt system demonstrated no signs of obstruction. Exchange for an entire new system with an intervening medium pressure valve was performed. In addition, 20 cc of peripheral venous blood was injected into the epidural L4–L5 space for a possible spinal leak as the cause of shunt malfunction. Neurologic improvement was seen and a repeat CT scan done the following day showed decreased ventricular size (Figure 1D). She was discharged home a few days later in an excellent neurologic condition, although resolution of the left-sided homonymous hemianopsia had not been seen yet. During follow-up, there were no complaints and she remained well, with gradual improvement of the visual field defect.

Eight months later, she presented to the emergency department with a right-sided parietal headache and visual blurring. Neurologic examination disclosed no abnormalities, and skin inspection showed neither redness nor swelling along the shunt path. CT scan of the head showed slit ventricles, and the presumed diagnosis was shunt overdrainage (Figure 2A). A shunt tap was performed and 03 cc of CSF was sent for routine examination, which was verified to be normal. She was sent home with a prescription for non-opioid analgesics as needed. Two days later, she returned to the emergency department with a mild fever (37.8°C/100°F), simple partial motor facial seizures, and mental confusion. An intravenous loading dose of phenytoin was administered. Reverse-transcription polymerase chain reaction testing for coronavirus disease 2019 was negative. MRI of the head demonstrated restricted diffusion in the right hippocampus, without enhancement, and slit ventricles. Blood test evaluation demonstrated hypokalemia and hypomagnesaemia. Although the most likely diagnosis was an unspecified viral systemic infection with electrolyte disturbances, intravenous acyclovir for possible herpes simplex encephalitis was initiated. A lumbar puncture was performed using a 25-gauge spinal needle and 10 cc of CSF was withdrawn. Routine examination showed 06 leukocytes/μL normal glucose, and elevated protein levels (120 mg/dL), consistent with a CSF block. CSF viral panel for herpes simplex, varicella-zoster virus, and herpesvirus-6.

Figure 1. (A) Admission T1-weighted magnetic resonance imaging with gadolinium showing a homogeneously enhancing pineal region mass (arrow), meningeal enhancement along the right cerebral convexity and slit ventricles. (B) Postoperative computed tomography (CT) scan of the head demonstrating pneumocephalus in the bifrontal regions and along the surgical corridor, but otherwise unchanged ventricular size. (C) CT scan of the head obtained 4 days after surgery/lumbar puncture when the patient complained of nausea, somnolence, and diplopia. Hydrocephalus can be seen. (D) CT scan of the head done the day following the shunt revision/blood patch showing improved ventricular size and near-complete resolution of the pneumocephalus.
was negative. Three days later, she complained of somnolence, unsteady gait, diplopia, and visual blurring. Neurologic examination disclosed bilateral sixth nerve paresis with horizontal gaze nystagmus and gait ataxia. A CT scan of the head showed significant hydrocephalus, along with marked enlargement of the perimedullary cisterns, consistent with a block at the foramen magnum, and distortion of the brainstem (Figure 2B). A shunt malfunction due to a lumbar CSF leak was believed to be the cause of the ventriculomegaly and neurologic deterioration. She was taken to the operating room and a lumbar epidural L4–L5 blood patch using 13 cc of peripheral venous blood was performed. A shunt tap disclosed a low pressure and free flow through the system was verified. A sample of 3 cc of CSF was withdrawn for examination. Immediately after the procedure the patient was more alert, conversing, and fully oriented. A CT scan of the head done 5 hours later showed a small decrease in ventricular size. A CT scan of the head done 3 days later showed marked improvement in ventricular size (Figure 2C). CSF results showed normal leukocyte count, glucose, and protein levels and were once again negative for herpes simplex, varicella-zoster virus, and herpesvirus-6. Bacterial cultures were negative in all CSF samples. Acyclovir was discontinued, and the patient was discharged home without any neurologic symptoms. On late follow-up, she was asymptomatic and neurologically intact.
DISCUSSION

LPH is a rare disorder in which there is ventricular dilation despite low or negative ICPs. Although most occurrences have been reported in patients with VP shunts, it may also be seen in non-shunted patients. LPH may either occur spontaneously or as a result of CSF egress from the skull base, spinal intradural procedures, or lumbar punctures.

Patients usually present with symptoms similar to those seen in the more prevalent high-pressure hydrocephalus. Headaches, nausea, cranial neuropathies, disturbances in mental status, and decreased consciousness are often seen. Ventriculomegaly can cause headaches in the setting of LPH even with very low pressures. Distension of the lateral and III ventricles may account for dysfunction of the association/projection fibers and hypothalamic dysfunction. While brainstem distortion may account for cranial nerve paresis, ataxia, nausea, vomiting, and decreased mentation.

In our patient, a complete block at the foramen magnum was evidenced in the imaging studies, with accumulation of CSF and enlargement of the perimedullary cisterns leading to brainstem compression, which explains the symptoms experienced. Shunt malfunction with LPH was seen first in our patient 4 days after a lumbar puncture was performed during a craniotomy. In this first episode, it is likely that the lumbar puncture was the reason for the development of low-pressure hydrocephalus, although aspiration of CSF during the craniotomy may have as well altered the CSF dynamics. The second occurrence of LPH, however, the only possible cause was the diagnostic lumbar puncture that had been performed 3 days earlier. The symptoms experienced before the lumbar puncture were probably related to a systemic viral illness with electrolyte abnormalities, which eventually resolved, and unlikely played a role in the development of the hydrocephalus. The restricted diffusion seen in the right hippocampus was likely a post-ictal abnormality, since no contrast enhancement was seen and CSF findings were not supportive of encephalitis. The immediate and sustained improvement in the patient’s symptoms as well as resolution of the ventriculomegaly following the blood patch confirmed that the lumbar puncture was the cause of the LPH.

The vast majority of shunted patients undergo lumbar punctures during their lifetime, although only a minor proportion develop LPH. Ventriculomegaly in the setting of LPH occurs when there is an alteration in brain compliance (elastance) as a predisposing factor. Radiation therapy, diffuse brain injury, low-flow ischemic states, and/or cerebral infarctions may alter brain viscoelastic properties, decrease brain turgor (Kb), and increase brain compliance. Low brain turgor allows more brain deformation and, when combined with an increase in the transmantine pressure, may lead to progressive ventricular dilation. Since our patient received radiation therapy at the age of 10 years, a low brain turgor likely played a role in the pathogenesis of LPH.

Dias et al. suggested that after a lumbar puncture there would be a decrease in the cerebral subarachnoid CSF leading to a progressive reduction in ICP to a point that it would prevent the valve from opening, leading to hydrocephalus. In contrast, a few arguments contest this theory. Many patients have a less than effective communication between the subarachnoid, ventricular, and spinal compartments.

The drop in ICP following a lumbar puncture is considerably smaller in those patients with a block at the cisterna magna, since the amount of CSF that actually passes through the obstruction is negligible. Barami et al. studied the pressure volume index in 20 shunted patients with hydrocephalus and demonstrated that the shift of cerebral subarachnoid CSF into the spinal canal is too small to account for the necessary drop in ICP needed to modify the CSF to venous pressures relationships. Therefore, in our patient, who had an overt obstruction at the level of the foramen magnum, it is unlikely that a considerable drop in ICP secondary to spinal CSF egress had occurred.

The pulsatile vector theory has also been proposed as an explanation for LPH. In chronically shunted patients, the CSF hydrodynamics are particularly altered since the shunt drains the CSF into a “third space” and there is no net state of CSF production and absorption, leading to persistent low ICP conditions and slit ventricles. This theory also holds the premise that a lumbar puncture will lead to loss of cerebral subarachnoid CSF, generating an increase in the interstitial fluid shockwaves with ventricular volume overload. ICPs remain low due to an outflow of the intracranial blood volume previously pooled in the venous system and a dampened ICP waveform coupled with an even lower ICP may prevent the shunt valve from opening, leading to hydrocephalus. Again, lack of CSF communication through the foramen magnum would not clearly explain the preferential intracranial subarachnoid CSF drainage as the initial pathophysiological mechanism in our patient.

The cerebral venous system is as equally important as the cerebral CSF circulation in the physiology of ICP control. The cortical cerebral venous pressure is slightly greater than the subarachnoid CSF and ventricular pressures under physiological conditions. On the other hand, the superior sagittal sinus and the deep periventricular veins, which are directly connected, have lower but equal pressures. In the supine position the cerebral venous drainage occurs predominantly through the jugular veins, whereas in the upright position it occurs through the spinal epidural venous system, which acts as a syphon directing the blood flow into the inferior vena cava system. Controlled outflow of cerebral venous blood is performed by the bridging veins, which act as a Starling resistor so as to prevent venous overdrainage in the upright position.

We postulate that in our patient the lumbar puncture lowered the spinal subarachnoid CSF pressure, particularly in the upright position, as leaking CSF was likely continuously aspirated into the negative-pressure epidural space. Since the spinal canal is a closed system, decreased CSF pressures led to even lower pressures in the spinal epidural venous system, increasing the syphoning effect and overdrainage of cerebral venous blood ensued. A decrease in periventricular deep veins pressures
led to an increased periventricular transparenchymal pressure gradient and ventricular dilation ensued in the setting of low brain turgor.\(^{1,11}\) The hypothesis that cerebral venous overdrainage has a role in the development of LPH in patients with shunts has already been proposed by Barami,\(^{1}\) but the correlation between venous overdrainage and a lumbar puncture had not been previously inferred.

Epidural blood patching has been classically used for post-lumbar puncture headaches, although how it works has not been fully elucidated yet.\(^{26,29}\) The indications for an epidural blood patch have become broader and now include spontaneous intracranial hypotension, post-lumbar puncture paradoxical herniation, treatment of cranial nerve dysfunctions and venous overdrainage in the pathogenesis of CSF pressure.\(^{2,25}\) This enabled an increase in the dural sinuses following lumbar puncture or spinal CSF diversion.

In our patient, we do not think the blood patch shifted the spinal CSF fluid into the intracranial space and led to an increase in the ICP, since there was no block at the foramen magnum. We believe that the epidural blood patch immediately increased the spinal epidural pressure, redirecting the venous flow and decreasing the cerebral venous overdrainage into the vena cava system.\(^{25}\) This enabled an increase in the cerebral venous blood volume and a rise in the deep cerebral veins pressure. Increasing the dural sinuses and periventricular veins pressures allows normalization of the periventricular transparenchymal pressure gradient and enables ventricular volume reduction, since the ventricles decrease in size only if their pressure is lower than the cerebral deep veins pressure.\(^{11,21}\) Later on, the blood patch most likely sealed the dural opening permanently, ultimately restoring the spinal CSF and venous pressures to a normal state, leading to a good long-term outcome.

CONCLUSIONS

Our case provides evidence that a lumbar blood patch may be successful in adult patients with post-lumbar puncture LPH, with good long-term results. Accumulation of CSF with enlargement of peri-medullary cisterns causing brainstem compression accounts for some of the symptoms encountered in this subset of patients who have CSF blockade at the foramen magnum. The role of cerebral venous overdrainage in the pathogenesis of LPH may be linked to variations in the spinal CSF pressure after a lumbar puncture.

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