Engorged epidural venous plexus and cervical myelopathy due to cerebrospinal fluid overdrainage: a rare complication of ventricular shunts

Case report

STACEY QUINTERO WOLFE, M.D., SANJIV BHATIA, M.D., BARTH GREEN, M.D., AND JOHN RAGHEB, M.D.

Division of Pediatric Neurosurgery, Department of Neurosurgery, University of Miami and Miami Children’s Hospital, Miami, Florida

✓ The authors report on a 17-year-old boy with cervical myelopathy from dilated epidural veins due to cerebrospinal fluid (CSF) overdrainage. The patient had a long-standing subdural–peritoneal shunt and presented with incapacitating spastic tetraparesis. Magnetic resonance imaging revealed significant cervical spinal cord compression from a markedly dilated epidural venous plexus. The shunt was externalized so that CSF flow dynamics could be assessed, and the patient was found to have low intracranial pressure (ICP). The patient was gradually acclimated to higher ICPs, and a new shunt was placed with an antisiphon device and a programmable valve set at the higher pressure. Postoperatively the child experienced significant clinical improvement, and reduction of spinal cord compression was evident on images. Compensatory engorgement of the epidural venous plexus due to long-term shunt usage should be considered in the differential diagnosis when cervical myelopathy due to a dilated epidural venous plexus is present.

KEY WORDS • cervical myelopathy • epidural venous plexus • ventriculoperitoneal shunt complication • intracranial hypotension • pediatric neurosurgery

Abbreviations used in this paper: CCJ = craniocervical junction; CSF = cerebrospinal fluid; CT = computed tomography; ICP = intracranial pressure; MR = magnetic resonance.
standing encephalomalacia around the right temporal cavity where the surgery had been performed. There was no change in ventricle size, and stable right brain shift was evident (Fig. 1A and B). No tumor recurrence was noted. Further MR imaging of the brain revealed diffuse dural enhancement (Fig. 1C and D) that was unchanged from previous imaging studies. Cervical MR imaging demonstrated significant spinal cord compression caused by a markedly dilated epidural venous plexus (Fig. 2). The spinal cord was triangular in shape, with large veins indenting the ventrolateral aspects and complete loss of the subarachnoid CSF space.

Angiography confirmed extreme dilation of the epidural veins with collapse of both internal jugular veins and engorged muscular and pericondylar veins shunting blood into the epidural venous plexus (Fig. 3). The decreased internal jugular venous drainage, relative to the caliber of the epidural venous plexus, raised the possibility of jugular stenosis. Stents were placed in the internal jugular veins bilaterally, without clinical improvement. The patient underwent multiple procedures for venous embolization but his condition continued to deteriorate. The epidural and intrathoracic venous pressures were measured and found to be within the normal range.

The performance of a shunt tap revealed no spontaneous flow, but easy withdrawal of CSF. Cerebrospinal fluid obtained from the tap showed no cells and protein moderately elevated at 118 mg/dl. Multiple lumbar puncture attempts revealed no spontaneous CSF flow (a “dry tap”). No evidence of a spinal CSF leak could be found on CT myelography.

Operation and Postoperative Course. In an effort to determine whether the child was dependent on his shunt or if his symptoms were caused by shunt overdrainage, the shunt was externalized to assess CSF flow dynamics. For the first 18 hours after externalization, there was no spontaneous CSF drainage although CSF could be aspirated. Initially, ICPs greater than 0 cm H₂O at the external auditory canal resulted in severe headaches and vomiting. Over the course of a week, the patient was gradually accustomed to higher ICPs by raising the height of the ventriculostomy drainage bag by several centimeters of H₂O daily. When his ICP was 15 cm H₂O, an MR image was obtained which showed a

---

**Fig. 1.** Axial CT (A) and T₁-weighted MR (B) images showing the right temporal surgical cavity with stable right brain shift and a flanged subdural-peritoneal shunt. C and D: Coronal T₁-weighted Gd-enhanced MR images showing diffuse dural enhancement, with characteristic lack of enhancement at the leptomeninges.

**Fig. 2.** Cervical MR images demonstrating significant spinal cord compression from a markedly dilated epidural venous plexus. Sagittal T₁-weighted Gd-enhanced (A) and T₂-weighted (B) images showing flow voids, indicating high flow and complete loss of subarachnoid CSF. Enhanced T₁-weighted images (C and D) showing deformation of the spinal cord with large veins indenting the ventrolateral aspects.
Cervical myelopathy due to CSF overdrainage

![Image 1](image1.png)

Fig. 3. Lateral (left) and anteroposterior (right) angiograms demonstrating extreme dilation of the epidural venous plexus and relative stenosis of the internal jugular veins bilaterally due to preferential drainage through the engorged epidural veins.

A dramatic reduction in the epidural vein volume with relief of cord compression and the reestablishment of the subarachnoid space, although myelomalacia with decreased spinal cord caliber was noted (Fig. 4). Clinically, the patient still had significant weakness, but less spasticity. The patient was taken to surgery where a medium pressure valve without an antisiphon device was changed to a programmable Strata valve set at 2.0.

The patient began an aggressive rehabilitation program, and after 3 months his strength had improved remarkably and he could walk again. The patient’s condition has continued to improve since his surgery. At 1.5 years since the shunt revision, his condition has returned to baseline in everything except fine motor skills, which have shown only mild improvement. Follow-up imaging results remain stable and show no spinal cord compression.

![Image 2](image2.png)

Fig. 4. Midline sagittal (A) and axial (B and C) T2-weighted MR images obtained 1 week after shunt externalization and return to normal ICP. A dramatic reduction in the size of the epidural veins, a reestablished subarachnoid space, and the resolution of cord deformation can be seen. Myelomalacia is present from chronic cord compression.

Discussion

Cervical myelopathy due to engorged epidural veins from CSF overdrainage is a rare shunt complication that arises due to a complicated interplay of fluid dynamics and unique craniospinal anatomy. In an enclosed system like the dura mater–covered neuraxis, hydrodynamic changes can affect the complicated venous anatomy of the CCJ, causing engorgement of the epidural venous plexus and subsequent spinal cord compression.

Pathophysiology of Symptoms

Intracranial hypotension results from CSF volume depletion, which may be caused by a hypovolemic state (such as dehydration), spontaneous CSF leak (usually due to a meningeal diverticulum), cranial or spinal trauma with CSF leakage, postoperative CSF leakage, or shunt overdrainage.

Patients with intracranial hypotension may present with multiple symptoms (neck pain; emesis; dizziness; auditory or visual changes; superior binasal visual field cut; third, fourth, and sixth cranial nerve palsies; bulbar weakness; labyrinthine hydrops; galactorrhea; interscapular pain; radiculopathies; parkinsonism; ataxia; frontotemporal dementia; encephalopathy; and coma), but headache is by far the most common symptom. The headaches associated with intracranial hypotension are thought to be due to the descent of the brain after a loss of CSF buoyancy, which causes traction on pain-sensitive structures such as the cerebral veins and arteries. This explains the postural nature of these orthostatic headaches and the patient’s relief when recumbent. Supratentorial and tonsillar descent of the brain can be seen on MR images obtained in symptomatic patients with intracranial hypotension.

Cerebrospinal fluid dynamics must be understood in light of the Monro–Kellie doctrine. With decreased CSF volume...
and a relatively constant brain volume, compensation occurs with an increase in blood volume.11 This increase is reflected primarily in the venous system and results in diffuse meningeal venous hyperemia and engorgement of the venous sinuses and pituitary gland, which can be seen on MR imaging as dural (but not leptomeningeal) enhancement.15 If these changes are insufficient to compensate for the loss of CSF, space-occupying lesions (such as subdural effusions or hematomas) may develop to maintain the stability of the intracranial volume.

Compensatory changes can also take place at the spinal level (Fig. 5). The vertebral canal is encased by bony and fibrous components that are minimally elastic. Unlike the cranial cavity, in which the dura is closely applied to the bone, the spinal canal has an epidural space in which lies the epidural venous plexus. The epidural venous plexus is a valveless plexiform network with a longitudinal pattern; it connects with the cranial venous drainage superiorly and continues inferiorly along the spinal cord as described by Batson in 1957.1 The venous drainage of the CCJ is anatomically diverse, but is generally referred to as the internal vertebral venous plexus.5 The anterior venous drainage is formed by the anterior condylar vein, which communicates with the cranial venous drainage and is the most consistent portion of the plexus.17 There may also be connections to the basilar plexus, and laterally there are connections with the vertebral artery venous plexus.17 The posterior internal vertebral venous plexus is less developed and extends over the C2–3 region.17 The plexus is further complicated by a confluence of the mastoid emissary veins, which unite jugular and internal vertebral venous systems, and the posterior condylar veins, which arise at the level of the jugular foramen.3

Subarachnoid collapse in response to CSF volume depletion can result in compensatory engorgement of this epidural venous plexus.11 Förderreuther et al.,4 reported on eight patients with orthostatic headaches in whom prominent, convex, dilated internal vertebral venous plexi were demonstrated on MR imaging; however, the patients they described did not manifest any symptoms of spinal cord compression. When venous engorgement becomes chronic, fibrocollagenous proliferation contributes to dural thickening,13 which may explain why what begins as compensatory venous stasis can progress to spinal cord compression. In addition to engorgement of the internal vertebral venous plexus, spinal MR imaging in the setting of intracranial hypotension may also demonstrate diffuse dural enhancement, similar to that in the brain, due to venous hyperemia.9 Extravasated fluid collections may be seen,16 but are usually due to extravasated CSF from meningeal defects in spontaneous intracranial hypotension, which must be differentiated from the scenario presented here.

In the literature, there is only one other case of cervical myelopathy from epidural venous engorgement due to shunt overdrainage. Miyazaki and colleagues8 described a case in which a patient who underwent placement of a ventriculoperitoneal shunt with a low-pressure valve suffered subarachnoid hemorrhage presented 15 months later with symptoms of mild myelopathy. A lumbar puncture produced CSF only with aspiration, and the results of MR imaging of the CCJ were similar to those in our patient: dilated epidural veins, loss of subarachnoid space, and spinal cord deformation. There was significant clinical and imaging-documented improvement after shunt ligation. Also similar to our case, the patient had no headache despite markedly low ICP. Although unusual, this phenomenon has been previously reported.15

Matsumoto and coworkers13 described the case of one other patient who presented with cervical spinal cord compression after ventriculoperitoneal shunt placement. Their case differs from our own in that the cord compression was due to an enhancing thickened dural mass, which may have been exuberant dural hyperemia, rather than to engorged veins. Additionally, their patient had a normal ICP, although there was resolution of the symptoms and the mass after shunt removal.

Case Evaluation

The present case raises several points from which we can learn. First, cervical myelopathy as a complication of CSF shunt overdrainage in the absence of the classic symptoms was unfamiliar to us and led to a significant delay in proper treatment. Because the shunt was long-standing, without recent manipulation or changes on imaging, and a shunt tap...
Cervical myelopathy due to CSF overdrainage

readily produced CSF, the possibility of the shunt as a cause was initially discarded. The absence of headaches also led us away from this diagnosis.

Because progressive spinal cord compression and myelopathy with an obvious venous abnormality were present, we began by treating the effect rather than the cause of the problem. After many failed treatments and worsening symptoms, the shunt was externalized to determine its involvement. Only then did the pathophysiology of the symptom complex become clear. The shunt’s involvement was further confirmed with the resolution of venous engorgement following the reestablishment of normal intracranial fluid dynamics. In retrospect, the MR images of the brain had shown diffuse dural enhancement for several years, suggestive of intracranial hypotension. The dry lumbar punctures were another clue to the presence of intracranial hypotension, although this finding is not entirely uncommon in patients who have had shunts for a long time.

Of note, this patient underwent radiotherapy after a subtotal resection of a pilocytic astrocytoma. Although it is unclear if this was in response to further growth of the residual tumor or as part of the primary treatment, our current practice is to avoid using irradiation in a child this young, opting instead for repeated resection or adjuvant chemotherapy when there is evidence of tumor progression.

Treatment and Prevention

Suspecting intracranial hypotension as a cause of venous plexus engorgement is the most important step in treating this entity. Cerebrospinal fluid dynamics can then be assessed using a shunt tap (which may be difficult due to the low ICP) or by externalizing the shunt. The reestablishment of normal ICP is of paramount importance. Shunt removal or ligation is optimal if the patient is not shunt dependent, but it is unlikely that patients with long-term shunt dependence can tolerate shunt occlusion. Increasing the ICP by gradually raising the external ventriculostomy drainage bag worked well for our patient, although he experienced marked discomfort and required close observation during the process. We placed a programmable valve so that we could continue to change the pressure as needed. Prevention of this rare phenomenon in patients with shunts may be accomplished with the use of medium- or high-pressure valves with antisiphon devices.

Conclusions

The shift of fluid between the cranial and spinal compartments and between the vascular and extravascular compartments keeps the total volume of the neuraxis constant and prevents dramatic reductions in intracranial volume. Intracranial hypotension due to CSF shunt overdrainage can cause the rare but devastating complication of cervical myelopathy from an engorged epidural venous plexus. Chronic transference of low or negative ICP to the cervical subarachnoid space may result in compensatory paravertebral venous engorgement which can deform the cord with resulting tetraparesis and myelomalacia. These effects can occur in a long-standing shunt even without the classic signs and symptoms of shunt overdrainage. Although it is extremely uncommon, the presence of prominent epidural venous channels should alert the physician to the diagnosis of intracranial hypotension. Reversal of intracranial hypotension can be accomplished with surgical revision of the shunt to include an antisiphon device.

References


Address reprint requests to: John Ragheb, M.D., Department of Neurosurgery, University of Miami, 1095 Northwest 14th Terrace, Miami, Florida 33136. email: jragheb@med.miami.edu.