Brain Neurorehabil. 2023 Nov;16(3):e27
https://doi.org/10.12786/bn.2023.16.e27
pISSN 1976-8753·eISSN 2383-9910

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Brain & NeuroRehabilitation

Case Report

Cervical Myelopathy Due to Epidural Hematoma at the Cervicomedullary Junction Associated With Ventriculoperitoneal Shunt Overdrainage: A Case Report

Ji Woong Park, Junwon Park

Received: Feb 14, 2023
Revised: Jul 28, 2023
Accepted: Aug 4, 2023
Published online: Oct 26, 2023

Correspondence to
Junwon Park
Department of Physical Medicine and Rehabilitation, Soonchunhyang University Seoul Hospital, Soonchunhyang University College of Medicine, 59 Daesagwan-ro, Yongsan-gu, Seoul 04401, Korea.
Email: jw910323@gmail.com

HIGHLIGHTS

- Ventriculoperitoneal (V-P) shunt placement is a frequently performed treatment.
- This is a case of myelopathy due to epidural hematoma by V-P shunt overdrainage.

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OPEN ACCESS

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Email: jw910323@gmail.com

ORCID iDs
Ji Woong Park
https://orcid.org/0000-0003-1680-0048
Junwon Park
https://orcid.org/0000-0001-7987-3581

Funding
This research was supported by the Soonchunhyang University Research Fund.

Conflict of Interest
The authors have no potential conflicts of interest to disclose.

ABSTRACT

We present a case of cervical myelopathy caused by epidural hematoma formation due to chronic cerebrospinal fluid overdrainage. A 55-year-old man who underwent ventriculoperitoneal (V-P) shunt surgery for normal pressure hydrocephalus presented with progressive weakness of both the upper and lower extremities. Magnetic resonance imaging (MRI) revealed compressive myelopathy at the cervicomedullary junction at the C1–C2 level caused by epidural hematoma formation due to intracranial hypotension (IH) caused by a complication of V-P shunt. He underwent decompressive laminectomy and hematoma removal at C1–C2 and replacement of the V-P shunt valve. Follow-up cervical spine MRI showed an improved state of severe central spinal stenosis at the C1–C2 level and an improved state of compression-related cord signal intensity change in the spinal cord. After surgical intervention and intensive rehabilitation, the patient showed clinical improvement. If cervical myelopathy is suspected in patients with a shunt, cord compression due to venous engorgement or hematoma caused by over-shunting and IH should be considered.

Keywords: Ventriculoperitoneal Shunt; Intracranial Hypotension; Spinal Cord Diseases

INTRODUCTION

Shunt placement is a frequently performed procedure for different pathologies, including hydrocephalus, despite many known complications, such as infection and malfunction [1,2]. Cerebrospinal fluid (CSF) overdrainage is a well-known complication that can cause intracranial hypotension (IH) [2]. Patients with IH typically have postural headaches with one or more of the following symptoms: nausea, vomiting, dizziness, diplopia, neck pain, and blurred vision [3]. Myelopathy is a rare complication of IH due to CSF overdrainage where the cervical spinal cord is compressed by epidural venous plexus engorgement [3] or epidural hematoma. We report an unusual case of a patient with cervical myelopathy related to epidural hematoma formation at the cervicomedullary junction primarily due to IH associated with long-term CSF shunt overdrainage without any typical symptoms.

Author Contributions

Conceptualization: Park J, Park JW; Data curation: Park J, Park JW; Formal analysis: Park J, Park JW; Writing - original draft: Park J, Park JW; Writing - review & editing: Park J, Park JW.

CASE DESCRIPTION

A 55-year-old male patient presented with progressive gait disturbance and fine motor disorder for more than one month without any history of trauma. He had a history of craniotomy and aneurysm clipping due to subarachnoid hemorrhage caused by rupture of the right anterior communicating artery aneurysm 14 years ago. He underwent ventriculoperitoneal (V-P) shunt surgery for normal pressure hydrocephalus two months after craniotomy and aneurysm clipping. He had no other medical problems, such as diabetes, but had an 18 pack-years of smoking history. Because his gait disturbance had worsened recently, brain and cervical spine magnetic resonance imaging (MRI) was performed. Brain MRI showed no significant findings that could explain the patient's worsened symptoms. However, cervical spine MRI showed high vascularity and signal changes of the spinal cord at the cervicomedullary junction level (**Fig. 1**). Transfemoral cerebral angiography was performed. It showed no spinal arterio-venous fistula. He was transferred to the university hospital for further evaluation and management.

He complained of tingling sensation in both the upper and lower extremities. He also had neck pain but denied any postural headache. Motor function examination was carried out. According to the Medical Research Council (MRC) grading system, 5 means normal power. The right upper extremity was graded as 3 and his left upper extremity was graded as 2. Both lower extremities were assessed as MRC grade 3–4. He complained of hypesthesia and hypoalgesia below the C3 level, and it was graded as C2 ASIA D. He showed generalized hyperreflexia in all four extremities. Deep anal pressure sensation and voluntary anal contraction were preserved with a normal bulbocavernosus reflex. The initial Berg Balance Scale (BBS) score was 29, and the Spinal Cord Independence Measure (SCIM) score was 19.

He was able to perform outdoor gait and activities of daily living (ADL) independently before the onset of symptoms. However, he needed minimal assistance for indoor gait and moderate to maximal assistance for performing ADL, especially eating, dressing, and grooming.

Cervical spine MRI showed compressive myelopathy at the cervicomedullary junction at the C1–C2 level caused by epidural hematoma formation due to IH caused by a mechanical complication of V-P shunt (**Fig. 2**). MRI also showed left-side uncovertebral joint hypertrophy, left foraminal disc herniation at C4–C5 with left neural foraminal stenosis, and bilateral uncovertebral joint hypertrophy at C6–C7. Due to mechanical complications of V-P shunt, he

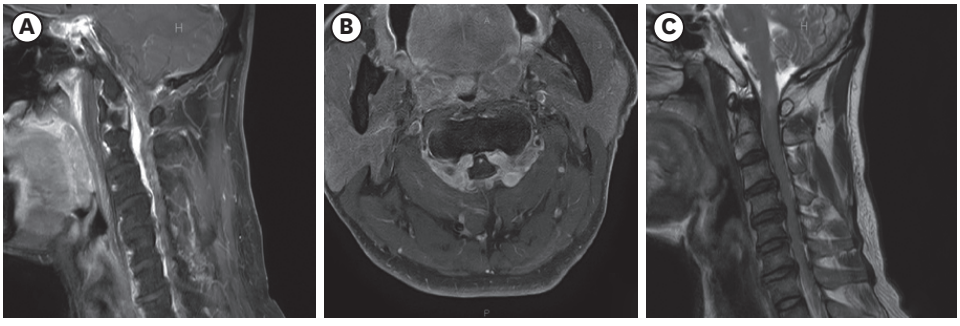

Fig. 1. Contrast-enhanced sagittal (A) and axial (B) T1 weighted MRI at the cervicomedullary junction at the C1–C2 level showing high vascularity and acute narrowing of the spinal canal and sagittal (C) T2 weighted MRI showing cord signal intensity change at the C2 level.
MRI, magnetic resonance imaging.

https://doi.org/10.12786/bn.2023.16.e27

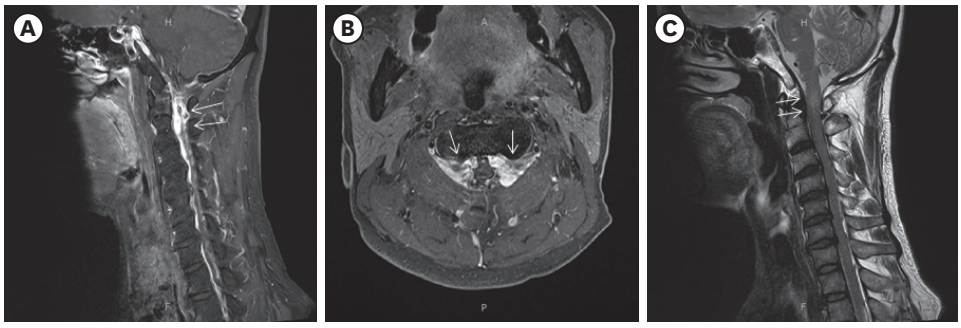

Fig. 2. Initial contrast-enhanced sagittal (A) and axial (B) T1 weighted MRI at the cervicomedullary junction at the C1–C2 level showing a space-occupying lesion in both lateral aspects of the spinal canal with high signal intensity and sagittal (C) T2 weighted MRI showing combined myelopathy at the cervicomedullary junction.
MRI, magnetic resonance imaging.

underwent decompressive laminectomy and hematoma removal at C1–C2 and replacement of the V-P shunt. The V-P shunt pressure was adjusted from 170 mmHg to 80 mmHg, and pulse steroid therapy was performed to promote early recovery and reduce the incidence of perioperative neurological complications [4].

Follow-up cervical spine MRI showed an improved state of compression-related cord signal intensity change in the spinal cord and severe central spinal stenosis at the C1–C2 level (**Fig. 3**). About one month later, decompressive laminectomy and posterior screw fixation at C3–C6 were carried out.

Electrodiagnostic examinations, including nerve conduction study (NCS), somatosensory and motor evoked potential (SEP and MEP) study, and needle electromyography (EMG) were performed. Sensory and motor NCS showed no abnormality. The SEP study showed mild dysfunction starting from both the upper extremities to the somatosensory cortex and moderate dysfunction starting from both the lower extremities to the somatosensory cortex. The MEP study showed moderate dysfunction starting from the motor cortex to both the upper extremities. Needle EMG showed increased insertional activity and abnormal spontaneous activity of the right pronator teres, extensor digitorum communis, left biceps, pronator teres, flexor carpi ulnaris, triceps deltoid, and bilateral paraspinalis (right C7, left C5–7 levels) muscles. The recruitment pattern was decreased for these muscles.

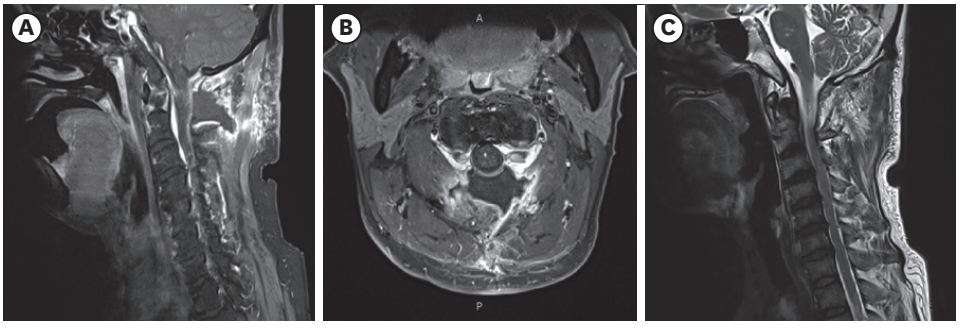

Fig. 3. Post-operative contrast-enhanced sagittal (A) and axial (B) T1 weighted MRI at the cervicomedullary junction at the C1–C2 level showing an improved state of a space-occupying lesion, especially in both lateral sides of the spinal canal. (C) Sagittal T2 weighted MRI showing a disappeared state of compression-related cord signal intensity change in the spinal cord.
MRI, magnetic resonance imaging.

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Table 1. Summary of the time course of symptoms and treatment

Time	Symptoms and treatment
2005	Craniotomy and aneurysm clipping, V-P shunt surgery
2019.11.	Onset of progressive gait disturbance and fine motor disorder, minimal assistance required for the walking indoor level
2019.12.28.	Decompressive laminectomy and hematoma removal at the C1–C2, and replacement of the V-P shunt
2019.12.28.–2020.1.5.	Pulse steroid therapy
2020.1.27.	Decompressive laminectomy and posterior screw fixation at the C3–C6
2020.1.27.–2020.2.3.	Pulse steroid therapy
2020.2.5.–2020.3.7.	Comprehensive rehabilitation therapy and supervision for the walking indoor level

V-P, ventriculoperitoneal.

Comprehensive rehabilitative treatment was carried out. In physical therapy, strengthening exercise for both the upper and lower extremities, dynamic standing balance training, and progressive gait training were performed. Occupational therapy was carried out to improve his hand functions impaired by cervical compression. ADL training sessions, including feeding and personal hygiene, were performed.

The patient's neurological condition gradually improved. The upper extremity motor power was improved to MRC grade 4 on the right side and grade 3 on the left side. For the lower extremity, muscle power was improved to MRC grade 4 on both sides. The BBS score was improved from 29 to 39, and the SCIM score was improved from 19 to 63. He was able to the walk indoor level under supervision. He was then discharged. A summary of the time course for symptoms and treatments is organized in a table (**Table 1**). This study was reviewed and approved by the Institutional Review Board (IRB) of Soonchunhyang University Seoul Hospital (SCHUH 2023-07-019). Informed consent was waived by the IRB.

DISCUSSION

Prompt diagnosis of cervical myelopathy caused by shunt complication can induce rapid recovery and good prognosis, as seen in this case. Cervical compressive myelopathy due to engorged epidural veins or epidural hematoma from CSF overdrainage is a rare shunt complication. Hydrodynamic changes can affect the complex venous anatomy of the cervicomedullary junction, causing engorgement of the epidural venous plexus and resulting in hematoma formation and subsequent spinal cord compression [5].

By diagnosing epidural hematoma, which is surgically correctable in a patient with shunt, the patient's clinical course could be improved. When patients with a shunt show progressive weakness or gait disturbance, evaluation of not only the brain but also spinal cord images should be performed. Further, compressive myelopathy caused by engorged epidural veins or epidural hematoma should be considered in the differential diagnosis of myelopathy in patients with V-P shunt [5].

In this case, the patient complained of neck pain without a postural headache. IH has a variety of causes with diverse manifestations. The cause is usually a leak of CSF, which can occur whenever the dura mater is damaged by a spinal surgery or trauma, lumbar puncture, craniotomy, or spontaneous injury. Shunt overdrainage might also be a cause of IH [3,6]. The main symptom of IH is postural headache. The absence of a headache is characteristic of overshunting-associated myelopathy [7]. Other common symptoms include nausea, vomiting, dizziness, neck pain, blurred vision, diplopia, tinnitus, ear fullness, and facial numbness [8].

https://doi.org/10.12786/bn.2023.16.e27

In the present case, engorgement of the spinal epidural venous plexus and pachymeningeal enhancement were observed, which are compatible with IH. Diffuse pachymeningeal enhancement over the cerebral hemispheres has been reported as the most characteristic neuroimaging finding of IH, although cases without any enhancement have also been reported [9]. Other described brain MRI findings are engorgement of venous sinuses, subdural fluid collections, pituitary enlargement, and downward displacement of the brain [9]. Extradural or subdural fluid collection, extradural extravasation of fluid, diffuse pachymeningeal enhancement, and engorgement of the spinal epidural venous plexus might be observed on spinal MRI [10].

In the present case, IH due to shunt overdrainage caused depletion of CSF, resulting in an epidural hematoma. Characteristic neuroimaging findings of IH are attributed to the Monro-Kellie doctrine; i.e., when the skull is intact, the sum of volumes of the brain, CSF, and intracranial blood remains the same [7]. Since the brain volume remains relatively constant, the decrease in the CSF volume is compensated primarily by an increase in the intracranial blood volume [2]. This increase is mainly reflected in the venous system, resulting in diffuse meningeal venous hyperemia, engorgement of the venous sinuses, and enlargement of the pituitary gland, which can be seen on MR imaging as dural enhancement [2]. If these changes are insufficient to compensate for the loss of CSF, space-occupying lesions, such as hematoma, may develop to maintain stability of the intracranial volume [2]. The Monro-Kellie doctrine might also be applied to the spinal canal because both the intracranial cavity and the spinal canal are closed systems [3].

In this case, the patient showed neural foraminal stenosis on cervical spine MRI. Generally, spinal canal stenosis is not reported as a cause of IH. However, a case has been reported in which the intracranial pressure increased due to spinal canal stenosis, resulting in a dural tear, which caused CSF leakage and ultimately caused IH. We should also consider spinal canal stenosis as an etiology of IH in patients with spinal canal stenosis who have a dural tear demonstrated by a cisternoscintigram or CT myelogram [11].

Although there might be no characteristic symptoms or imaging findings, suspicion of the possibility of IH as the cause of dilated epidural veins or epidural hematoma resulting in compressive myelopathy in a patient with a shunt is the most important step. The diagnosis of compressive myelopathy caused by epidural hematoma due to IH caused by a complication of V-P shunt was based on the history, physical examination, imaging study, and electrodiagnostic examination evidence. This was further supported by evidence of clinical and radiological improvement following replacement of the V-P shunt.

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