A case of suspected low-pressure hydrocephalus caused by spinal drainage following subarachnoid hemorrhage

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Abstract

Background: Hydrocephalus induced by low cerebrospinal fluid (CSF) pressure is extremely rare and sporadically reported. Subarachnoid hemorrhage, head trauma, and spinal drainage were reported to be the causative factors for surgical treatments.

Case Description: A 33-year-old male with subarachnoid hemorrhage caused by right vertebral artery aneurysm rupture developed a headache. A trapping surgery was performed, and a spinal drain was inserted from the lumbar L4/5 for subarachnoid hemorrhage washout. On postoperative day 3, increase in subdural fluid accumulation at the posterior fossa craniotomy site and narrowing of the cerebellar sulci appeared in addition to mild enlargement of the ventricles. The patient complained of a headache during head elevation. Low-pressure hydrocephalus (LPH) was suspected. The spinal drain was removed, the headache was relieved, and cerebral ventriculomegaly disappeared. The subsequent clinical course was good. The patient was discharged 3 weeks after the surgery.

Discussion: LPH is a rare disease caused by various factors and is treated by correcting liquorrhea or overdrainage, if any. Otherwise, drainage at a negative CSF pressure is necessary. Its symptoms and image findings are similar to those of intracranial hypertension and normal-pressure hydrocephalus. This paper reports a suspected LPH case caused by spinal drainage following subarachnoid hemorrhage with literature review.

Keywords: low-pressure hydrocephalus; low intracranial pressure hydrocephalus; negative-pressure hydrocephalus; subarachnoid hemorrhage; spinal drain

Introduction

Low-pressure hydrocephalus (LPH) is an extremely rare and sporadically reported disease. LPH was first proposed by Pang et al. in 1994. While its developmental mechanism is still unclear, subarachnoid hemorrhage, head trauma, posterior fossa surgery, lumbar puncture, and spinal drainage have been reported to be the causative factors for surgical treatments.

Some of its symptoms and image findings are similar to those of intracranial hypertension and normal-pressure hydrocephalus.² The symptoms include disturbance of consciousness, headache, vomiting, and cranial nerve palsy. The image findings are cerebral ventriculomegaly and periventricular edema (similar to intracranial hypertension). An LPH case due to spinal drainage following subarachnoid hemorrhage was encountered at Nippon Medical School Hospital, and this case is hereby reported.

Case description

A 33-year-old male patient with subarachnoid hemorrhage (World Federation of Neurosurgical Societies classification, grade II) caused by right vertebral artery fusiform aneurysm rupture developed a headache(**Fig. 1A, B, C**). The dissecting aneurysm lesion was located at a relatively lower position, almost about the height of the foramen magnum and proximal to the posterior inferior cerebellar artery. Fine branches were observed from the periphery of the aneurysm(**Fig. 1C, D**). Consequently, a craniotomy for trapping was performed. The surgical approach involved a suboccipital craniotomy with an atlas laminectomy. Moreover, the lesion was trapped with cerebral aneurysm clips(**Fig. 1E**).

Spinal drain was inserted from the lumbar L4/5 before surgery. Moreover, cerebrospinal fluid (CSF) drainage was continued after surgery for the washout of subarachnoid hemorrhage. On postoperative day (POD) 3, cerebral ventriculomegaly appeared, the subdural cavity at the posterior fossa craniotomy site was opened and fluid accumulation was observed, and the cerebellar sulci became narrow and tight(**Fig. 2**). In addition, the patient complained of a headache during head elevation on the same day. Low-pressure headache due to spinal drainage was suspected based on the headache induced by head elevation, the appearance of subdural fluid accumulation, and obscure sulci under spinal drainage(**Fig. 2**). Consequently, the spinal drain was removed, and the headache during head elevation was relieved promptly.

Furthermore, subdural fluid accumulation, obscure sulci, and cerebral ventriculomegaly improved as inferred from the images(**Fig. 2**).

The spinal drainage volumes were 46 and 55 mL on POD 1 and 2, respectively(**Fig. 3**). On POD 3, headache during head elevation appeared, and images showed cerebral ventriculomegaly and subdural fluid accumulation. Moreover, 71 mL of fluid had already been drained within half a day. The drain height was adjusted according to the drainage volume per hour with the target daily drainage volume of 100–120 mL. However, the drainage volume was small, and the drain was lowered to -7 cm using the external acoustic pore as the standard. Consequently, the cerebral ventricles were found to be enlarged despite the low CSF pressure. After drain removal, the headache was relieved promptly, and the CT findings improved(**Fig. 3**). Thus, the patient was diagnosed with LPH. Data of CSF when spinal drain was inserted: cell $68/\mu\text{L}$, protein 275mg/dL. That of CSF when spinal drain was removed: cell $435/\mu\text{L}$, protein 113mg/dL. The subsequent clinical course was good. No recurrence of cerebral vasospasm or hydrocephalus was noted. The patient was discharged 3 weeks after the surgery.

Discussion

LPH is expressed as negative-pressure hydrocephalus(NegPH) when the CSF pressure is

negative. LPH and NegPH are clinically distinguished from the other types of hydrocephalus. In addition, both types have rare characteristics that seem contradictory at a glance (i.e., cerebral ventriculomegaly in the state of low intracranial pressure). The symptoms include headache and disturbance of consciousness induced by low CSF pressure, dementia caused by cerebral ventriculomegaly, gait disturbance, and urinary incontinence. Many of the cases in which the CSF pressure was measured with lumbar puncture and intracranial pressure monitoring were reported to have a value <80 mmH₂O. Various causative factors and developmental mechanisms have been investigated. However, no definitive ones have been reported yet⁶.

The developmental mechanism of this case was examined in the study(**Fig. 4**). The CSF malabsorption caused by low CSF pressure resulting from excessive spinal drainage and compensatory intracranial hyperemia due to low CSF pressure may have occurred. Consequently, CSF accumulation and impaired CSF buffering capacity, leading to cerebral ventriculomegaly and hydrocephalus may have occurred. Furthermore, increased brain compliance due to subarachnoid hemorrhage and surgery may also have caused impaired CSF buffering capacity, contributing to cerebral ventriculomegaly. Furthermore, impaired CSF buffering capacity may have also occurred due to stenosis of the CSF tract due to cerebral ptosis caused by low CSF pressure⁹. Cutler et al.⁷ reported that CSF is not absorbed when the intracranial pressure is <68 mmH₂O, suggesting that CSF malabsorption was noted in this

case due to low CSF pressure as the drain was lowered to -7 cm using the external acoustic pore as the standard. According to the revised Monro–Kellie hypothesis, cerebral parenchyma + vascular bed + CSF cavity = constant⁸. Vascular bed and venous pressure increase in case of low CSF pressure, resulting in CSF malabsorption. In addition, stasis is also considered as a cause of hydrocephalus apart from malabsorption. Deformity and distortion of the CSF tract due to cerebral ptosis (an elliptical deformity of the pons, dropping of the splenium of the corpus callosum, and narrowing of the prepontine cistern) caused by low CSF pressure led to the impaired CSF outflow.⁵

Previously reported cases (19 cases) of LPH were reviewed in this study(**Table 1**). All cases developed LPH following surgical treatments for some kind of head disease or trauma. Liquorrhea repair was performed in one case in which postoperative liquorrhea was identified as the cause. In this case too, overdrainage due to spinal drainage was thought to be the cause. Thus, headache induced by low CSF pressure and image findings improved promptly after the drain was removed.

The causes of LPH in most cases were unclear despite low CSF pressure, liquorrhea, and overdrainage. However, LPH was thought to have developed due to intracranial diseases or craniotomy. In terms of the pathophysiology, Cheng et al.¹ and Hunn et al.⁴ have reported that extracellular fluid deficiency causes a change in the viscoelastic substance coefficient of the

cerebral parenchyma, leading to LPH. Treatment targeting hydrocephalus improvement by draining the CSF with the application of a pressure lower than the intracranial pressure or negative pressure with ventricular drainage has been reported for cases with no obvious liquorrhea.

In addition, CSF drainage was continued while increasing the intracranial pressure by lowering the head position and increasing the venous pressure by neck wrapping or by reducing blood sodium concentration through fluid replacement with a hypotonic solution in cases where the intracranial pressure became negative³.

Appropriate treatment for LPH may be delayed because its symptoms and image findings are similar to those of intracranial hypertension and normal-pressure hydrocephalus. Several LPH cases were difficult to diagnose and treat wherein multiple shunt reconstructions were performed based on the diagnosis of shunt failure because no improvement had been observed despite the reduction of shunt pressure to some extent with ventriculoperitoneal shunting. If cerebral ventriculomegaly does not improve even after shunt surgery for hydrocephalus, it is necessary to look for liquorrhea and consider the possibility of LPH. Moreover, the shorter the time from onset to the start of treatment, the better the post-treatment prognosis and the easier it is for ventricular size to revert to the original size. With appropriate treatment, the prognosis is good. Thus it is important to understand the pathology of LPH.

Conclusion

An LPH case due to spinal drainage following a subarachnoid hemorrhage surgery was encountered. It is necessary to consider LPH in cases with cerebral ventriculomegaly even if liquorrhea and overdrainage are suspected. Furthermore, the symptoms and image findings are similar to those of intracranial hypertension and normal-pressure hydrocephalus even though the condition might have been caused by intracranial diseases or craniotomy. Moreover, LPH has a good prognosis if diagnosed early and treated appropriately. Thus, it is important to consider the possibility of low CSF pressure when treating hydrocephalus. In addition, basic research is important for the elucidation of CSF physiology, which is still under discussion.

Conflict of Interest: The authors declare no conflicts of interest

References

1. Cheng Z, Wang W, Han Y, et al. Low pressure hydrocephalus: clinical manifestations, radiological characteristics, and treatment. *Br J Neurosurg.* 2017;31:410-4.
2. Filippidis AS, Kalani MY, Nakaji P, Rekate HL. Negative-pressure and low-pressure hydrocephalus: the role of cerebrospinal fluid leaks resulting from surgical approaches to the cranial base: Report of 3 cases. *J Neurosurg.* 2011;115:1031-7.
3. Pandey S, Jin Y, Gao L, Zhou CC. Negative-pressure hydrocephalus: a case report on successful treatment under intracranial pressure monitoring with bilateral ventriculoperitoneal shunts. *World Neurosurg.* 2017;99:812-e7.
4. Hunn BH, Mujic A, Sher I, et al. Successful treatment of negative pressure hydrocephalus using timely titrated external ventricular drainage: a case series. *Clin Neurol Neurosurg.* 2014;116:67-71.
5. Kim YS, Kim SH, Jung SH, et al. Brain stem herniation secondary to cerebrospinal fluid drainage in ruptured aneurysm surgery: a case report. *SpringerPlus.* 2016;5:247.
6. Wang X, Wang Z, Chen Y. Diagnosis and Treatment of Low Intracranial Pressure Hydrocephalus. *Int J Neurol Phys Ther.* 2017;3:35.
7. Cutler RW, Page L, Galicich J, Watters GV. Formation and absorption of cerebrospinal fluid in man. *Brain.* 1968;91:707-20.
8. Ota T. [Neurosurgery] 12th Revised Edition, Kinpodo, 2016, Japanese.

9. Mase M. [An Overview of Cerebrospinal Fluid Physiology: The Flow from the Classical Interpretation to the New Interpretation] *Currently Practical Neurosurgery*. 2018;28(7):663-7, Japanese.

Figure legends

Figure 1 Preoperative images and intraoperative findings of the case.

(A) (B) Brain computed tomography (CT) images of a 33-year-old male patient with subarachnoid hemorrhage caused by right vertebral artery fusiform cerebral aneurysm rupture.

(C) (D) Digital subtraction angiography (DSA) and 3D-DSA images. The lesion was located proximal to the posterior inferior cerebellar artery.

(E) A picture of trapping surgery.

Figure 2 The course of brain CT.

On POD 3, cerebral ventriculomegaly appeared, the subdural cavity at the posterior fossa craniotomy site was opened and fluid accumulation was observed, and the cerebellar sulci became narrow. After spinal drain removal (POD 11), subdural fluid accumulation, obscure sulci, and cerebral ventriculomegaly improved.

Figure 3 The vertical axis representing the drainage volume from the spinal drain on an hourly basis and CT findings arranged over time.

The drainage volumes from the spinal drain were 46 and 55 mL on POD 1 and 2, respectively. Moreover, 71 mL of fluid was already drained within half a day on POD 3 when headache during head elevation, cerebral ventriculomegaly, and subdural fluid accumulation appeared.

On POD 3, the spinal drain was removed, and the CT findings improved.

Figure 4 Schema of the mechanism of onset.

Cerebrospinal fluid malabsorption due to low CSF pressure and compensatory intracranial hyperemia caused by low CSF pressure occurred, resulting in CSF accumulation and impaired CSF buffering capacity, leading to cerebral ventriculomegaly and hydrocephalus. In addition, increased brain compliance due to subarachnoid hemorrhage and surgery also caused impaired CSF buffering capacity. Furthermore, impaired CSF buffering capacity occurred as a result of stenosis of the CSF tract due to cerebral ptosis caused by low CSF pressure.

Table 1 A summary of the previously reported LPH cases (19 cases).

Figure 1

Figure 2

Figure 3

Drainage volume from spinal drain (mL/h)

Figure 4

Table 1. A summary of the previously reported LPH cases (19 cases)

Author, year of publication	Age/Gender	Underlying disease	Affected surgical treatment	Treatment	GOS at discharge
Cheng, 2017	23/M	Acute subdural hematoma	Craniotomy for hematoma evacuation	Negative-pressure CSF drainage, head elevation	5
	41/F	Subarachnoid hemorrhage	Craniotomy clipping	Negative-pressure CSF drainage, head elevation	5
	42/M	Brain contusion	Craniotomy	Negative-pressure CSF drainage, head elevation	3
	61/F	Hypertensive cerebral hemorrhage	Craniotomy for hematoma evacuation	Negative-pressure CSF drainage, head elevation	4
	62/M	Brain contusion	Craniotomy	Negative-pressure CSF drainage, head elevation	5
	8/F	Astrocytoma	Tumorectomy via craniotomy	Negative-pressure CSF drainage, head elevation	5
	39/M	Brain contusion	Craniotomy	Negative-pressure CSF drainage, head elevation	5
Pandey, 2017	26/M	Epidermoid tumor (posterior fossa)	Endoscopic transnasal transclival tumorectomy	Neck wrapping, CSF drainage 200 mL/day	5
Hunn,2014	27/M	Pineal tumor	Craniotomy for extirpation	Negative-pressure CSF drainage, 120–360 mL/day	N/A
	51/M	Cavernous hemangioma	Craniotomy for extirpation	Negative-pressure CSF drainage, 120–360 mL/day	N/A
	65/F	Metastatic brain tumor	Craniotomy for extirpation	Negative-pressure CSF drainage, 120–360 mL/day	3
	12/M	Severe head trauma	Craniotomy	Negative-pressure CSF drainage, 120–360 mL/day	N/A
	36/M	Severe head trauma	Craniotomy	Negative-pressure CSF drainage, 120–360 mL/day	N/A
	54/F	Subarachnoid hemorrhage	Craniotomy clipping	Negative-pressure CSF drainage, 120–360 mL/day	N/A
	70/F	Unruptured cerebral aneurysm (MCA)	Craniotomy clipping	Negative-pressure CSF drainage, 120–360 mL/day	1
	63/M	Subarachnoid hemorrhage	Craniotomy clipping	Negative-pressure CSF drainage, 120–360 mL/day	4
Filippidis,2011	61/M	Subarachnoid hemorrhage	Craniotomy clipping	Negative-pressure CSF drainage, neck wrapping	5
	67/M	Cavernous vascular malformation, dural arteriovenous Fistula	Embolectomy, γ knife, resection, decompressive craniectomy for postoperative cerebellar infarction	Liquorrhea repair, third ventriculostomy	3
	56/F	Dissecting aneurysm	Craniotomy clipping	CSF drainage, liquorrhea repair	5