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Case Report

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Acute Onset of Syndrome of the Trephined After Lumboperitoneal Shunt Placement: A Case Report

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Conflict of Interest
The authors have no financial conflicts of interest.

ABSTRACT

Decompressive craniectomy is widely recognized as a life-saving emergency operation for the treatment of increased intracranial pressure; however, it can lead to severe complications, such as "syndrome of the trephined." Cerebrospinal fluid diversion, particularly after lumboperitoneal shunting, can affect the occurrence of this disease and worsen the symptoms. We report an acute case of this syndrome after lumboperitoneal shunting in a patient who had previously undergone decompressive craniectomy. The patient rapidly fell from a Glasgow Coma Scale (GCS) of 14 to a comatose state and a GCS of 4 only in 2 days. After cranioplasty, the patient recovered fully; however, this took a prolonged period.

Keywords: Decompressive craniectomy; Cerebrospinal fluid shunt; Hydrocephalus; Craniotomy; Intracranial hypotension

INTRODUCTION

The "Syndrome of the Trephined" (SoT), also known as the syndrome of sinking skin flap, is a condition in which a patient experiences neurological decline following a decompressive craniectomy, which can be reduced or relieved after cranioplasty.[8] Its clinical presentations vary and include headache, mild cognitive dysfunction, motor weakness, and mental deterioration, which can be as severe as a comatose state and death.[2,9] "Paradoxical herniation" can also manifest in the progress of SoT, where brain herniation occurs without external brain compression.[9]

SoT can be difficult to detect owing to the lack of specific symptoms and the masking of previous neurological sequelae. Acute SoT onset after cerebrospinal fluid (CSF) diversion may occur,[6,14] and lumbar-peritoneal (L-P) shunts might be more fatal than ventricular-peritoneal shunts because of their mechanism of action. Early detection and proper management may reduce permanent sequelae. However, there are only few case reports with even smaller amount of detailed treatment progress.[4,9] Here, we present the clinical course of the occurrence and treatment of acute SoT onset after the implementation of an L-P shunt procedure.

CASE REPORT

The authors state that the patient provided informed consent for the study by verbal consent. Furthermore, the authors declare that all work was carried out in compliance with the Ethical Principles for Medical Research Involving Human Subjects outlined in the Helsinki Declaration in 1975 (revised in 2000). This study was approved by the Institutional Review Board of the Busan Paik Hospital (IRB No. 2022-11-065).

Clinical course before the SoT

A 46-year-old woman presented to our hospital with drowsiness following a seizure. An initial brain computed tomography (CT) scan revealed a large intracerebral hematoma (ICH) in the left temporoparietal lobe, along with a mass effect compressing the ventricular system and shifting the midline toward the right side. This may have resulted from a cerebral venous sinus thrombosis. Brain magnetic resonance imaging showed an empty delta sign in the left transverse sigmoid sinus (**FIGURE 1A**). Although she promptly managed to control the increased intracranial pressure while being monitored in the emergency room, her mental state deteriorated to stupor after a few hours (**FIGURE 1B**). Immediate decompressive craniectomy of the left hemisphere was performed, followed by a successful clinical outcome (**FIGURE 1C**). Following magnetic resonance venography, we confirmed that her ICH was caused by cerebral venous sinus thrombosis in the left transverse sigmoid sinus. After a month, she was capable of self-ambulation with mild right hemiparesis and alert mentality with mild motor dysphagia, as assessed using a modified Rankin Scale score of 2.

Three months after the initial surgery, the patient's neurological status was maintained. However, she required rehospitalization because her external hydrocephalus worsened (**FIGURE 2A**). Initially, needle aspiration on the affected side and compression dressing were performed; however, the patient relapsed shortly thereafter. Thus, we planned a staged surgical treatment for CSF diversion and cranioplasty. Owing to the relatively collapsed ventricle size, we chose to use an L-P shunt for CSF diversion.

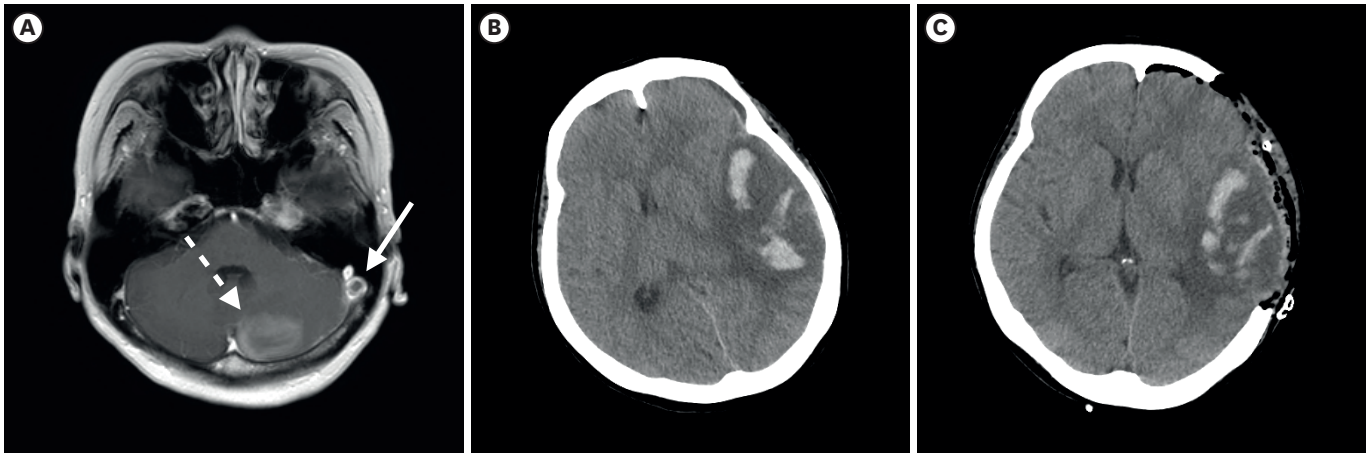

FIGURE 1. Initial preoperative and postoperative brain image following craniectomy. (A) Axial image of contrast enhanced brain magnetic resonance imaging shows an empty delta sign in left sigmoid sinus (white arrow) and acute infarction in the medial side of the left cerebellum (white dotted arrow). (B) A brain CT scan shows a large intracerebral hematoma in the left temporo-parietal lobe along with mass effect and midline shifting toward the right side. (C) A brain CT after craniectomy shows restoration of the midline shifting.
CT: computed tomography.

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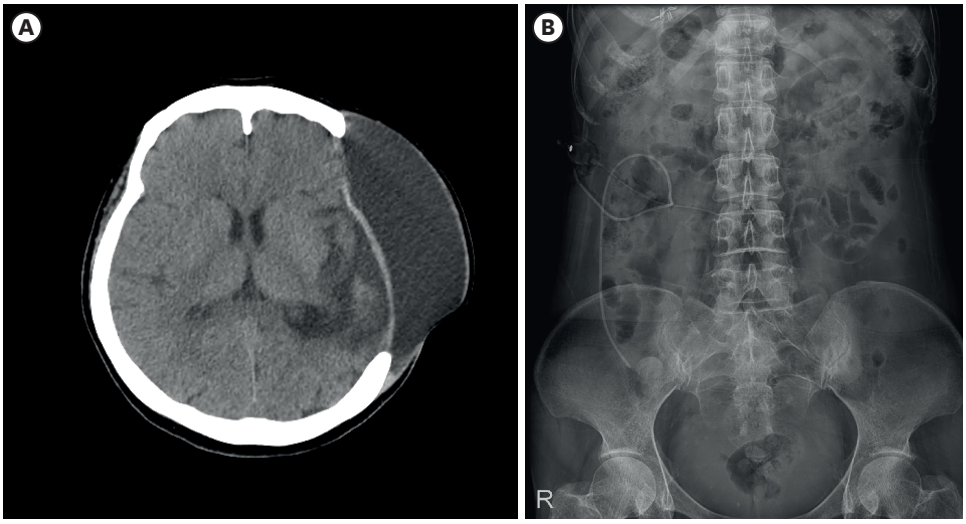

FIGURE 2. Brain image before lumboperitoneal shunt and postoperative X-ray. (A) Brain computed tomography after three months of craniectomy shows external hydrocephalus in the left hemisphere. (B) A simple abdomen plain X-ray anteroposterior view shows the lumboperitoneal shunt (proximal catheter placed between L3 and L4).

Lumboperitoneal shunt procedure

The L-P shunt was performed using the Strata NSC lumboperitoneal valve system (Medtronic, Minneapolis, MN, USA) without any issues (**FIGURE 2B**). The patient was placed in the right lateral decubitus position under general endotracheal anesthesia. A 14G thin Tuohy spinal needle was inserted into the subarachnoid space through a linear skin incision in the L3/4 interspinous space. The lumbar catheter was then inserted about 8 cm deep and brought into the flank from the lumbar area using a tissue tunneler. Next, a transverse linear skin incision was made in the right periumbilical area of the abdomen, and the peritoneum was carefully incised to pass the peritoneal catheter through the tunnel from the flank to the abdominal wound. After assembling and inserting the lumbar and peritoneal catheters into the fatty layer of the flank, the peritoneal catheter was inserted into the peritoneal cavity and fastened loosely with chromic. The surgical wound was then closed layer-by-layer.

Clinical course after the SoT

On the day after the L-P shunt operation, the patient experienced severe headaches, and her mental state deteriorated to drowsiness. On postoperative day (POD) 2, the patient's mental state worsened to a semicomatose state, her pupils were constricted, and showed no light reflex. Furthermore, her vital signs were unstable. CT images taken immediately (**FIGURE 3A**) showed that the ventricles were rapidly compressed, with the midline shifting toward the opposite side of the craniectomy site, implying a paradoxical herniation.

We promptly increased the valve-opening pressure to the maximum level, but there were no neurological changes. On POD 3, another surgery was performed to clamp the shunt and prevent further CSF drainage. However, the patient's semicomatose state persisted for the next three days, with no light reflex observed (**FIGURE 3B**). On POD 6, early cranioplasty was performed with thorough duroplasty using prolene to suture some CSF leakage points seen in the operation field, and the artificial dura was patched and covered with autologous bone. The next day, the light reflex promptly returned, and the patient's tachypnea subsided (**FIGURE 3C**).

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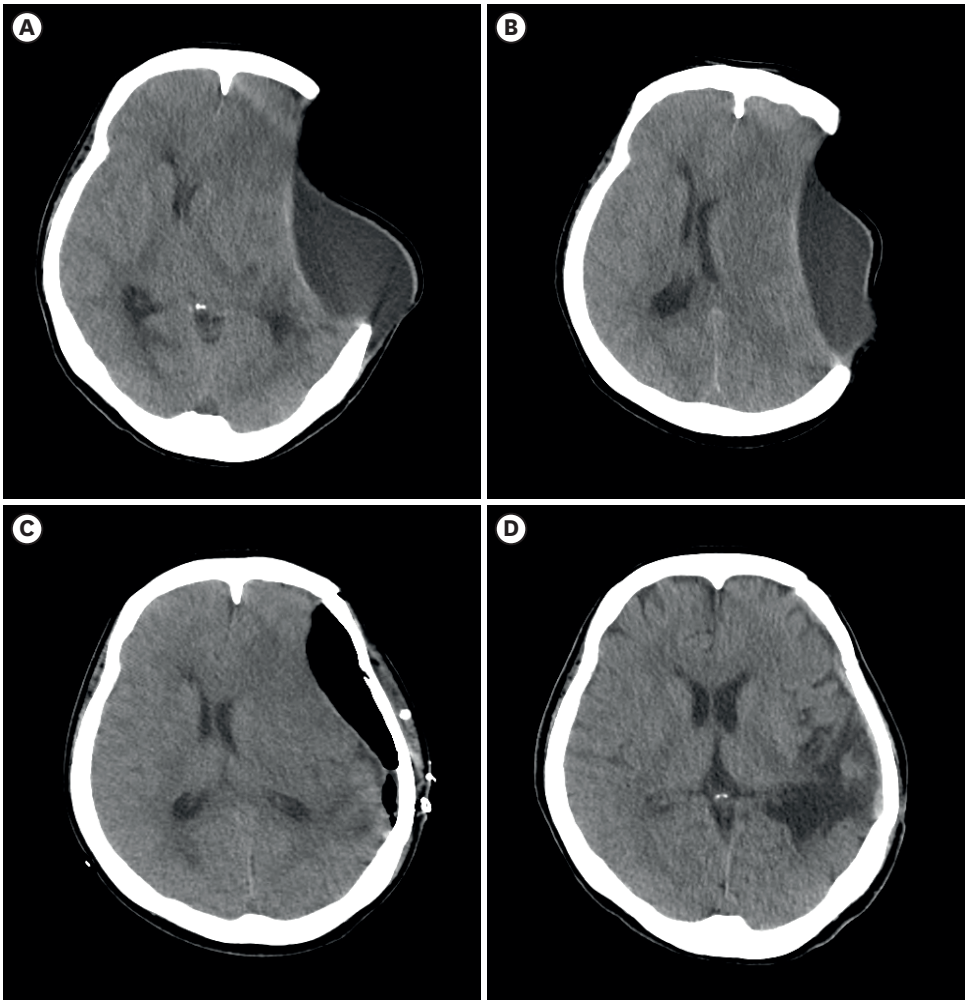

FIGURE 3. Preoperative and postoperative brain image following cranioplasty. (A) A brain CT from the second day of the shunt shows a sunken extradural cyst, ventricle, and midline shifting to opposite side. (B) A brain CT from the 6th day of the shunt shows a slightly increased ventricle, but the midline shifting is still visible. (C) The midline shifting is more improved after cranioplasty. (D) A brain CT after 3 months of cranioplasty shows completely recovered paradoxical herniation and external hydrocephalus.
CT: computed tomography.

A week later, she recovered from drowsiness and could cooperate, so she was moved to the general ward and was dismissed from the intensive care unit. Both arm motors improved first, and after less than two months, as both legs were better off, she could walk with minimal assistance. A few days later, a swallowing test was performed. The patient was free of L-tubes and started feeding independently. She was then transferred to the Department of Rehabilitation Medicine and discharged after another one month. Subsequently, the patient exhibited a slow but persistent recovery. Eventually, she fully recovered with minimal neurological symptoms, to the extent that she was able to walk into the clinic herself and even able to logically complain about her prolonged hospitalization (**FIGURE 3D**).

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DISCUSSION

Although the pathophysiology of SoT remains unclear, various studies have suggested theories to explain its development, including atmospheric pressure, abnormalities in CSF flow, dysfunction in cerebral metabolism, and decreased cerebral blood flow.[4,5,13] Conservative treatment options, such as intravenous hydration, maintenance of the Trendelenburg position, and avoidance of hyperosmotic agents, are available; however, these are often ineffective. Cranioplasty is considered the definitive treatment for SoT.[4,7]

In the present case, the patient developed SoT following L-P shunt placement and experienced an acute course. When the patient became semicomatose, attempts were made to increase the shunt valve pressure and ligate the L-P shunt. However, despite these conservative measures, there was no improvement in the patient's condition, and she experienced further complications. As a last resort, emergent cranioplasty was performed, which resulted in complete recovery.

SoTs associated with CSF diversion may be dangerous. However, there have been no systematic studies on this topic, and only a few case reports exist. To our best knowledge, the present case is a first SoT case combined with L-P shunt. We summarized SoT cases with CSF diversion of ventriculoperitoneal (V-P) shunt and lumbar spinal puncture in **TABLE 1**. In those cases, CSF diversion procedure, either of V-P shunt or CSF drainage through lumbar spinal puncture were performed during rehabilitation period when the patient was in stable status so it is able to detect onset time, symptom progress and recovery.[3,4,6,7,9,10,13-15]

TABLE 1. Summarized case reports regarding acute onset syndrome of the trephined following CSF diversion

Authors	Year	Sex	Age	Predisposing disease	CSF diversion	Onset of SoT	Treatment	Symptoms	Outcome
Kim et al.[10]	2020	Female	17	Traumatic subarachnoid hemorrhage, brain abscess	V-P shunt	Several months	V-P shunt clamping operation	Vomiting, ptosis, decreased eye contact	Recover
Chalouhi et al.[3]	2012	Female	66	Subdural hematoma, postoperative infection	V-P shunt	A week	Cranioplasty with titanium plate	Status epilepticus, decreased level of consciousness	Recover
Sakamoto et al.[13]	2005	Female	57	Aneurysmal subarachnoid hemorrhage	V-P shunt	2 years	Cranioplasty with titanium plate	Hemiparesis and confusion	Recover
Han et al.[7]	2008	Male	37	Traumatic subdural hematoma	V-P shunt	1 day	Cranioplasty with autobone flap	Decreased level of consciousness	Recover
Gschwind et al.[6]	2012	-	65	MCA infarction	CSF leakage after lumbar puncture	Several days	Epidural blood patch	Headache, vomiting, anisocoria, decreased level of consciousness	Recover
Dillen et al.[4]	2018	Male	25	Traumatic subdural hematoma, postoperative infection	V-P shunt	3 days	Plaster molded to patient's head with vacuum assistance	Decreased level of consciousness	Survive
Shen et al.[15]	2017	Male	29	Traumatic subdural hematoma	CSF drain (30 mL) with lumbar spinal puncture	1 day	Conservative treatment	Decreased level of consciousness (coma)	Recover
		Male	56	Traumatic subdural hematoma	CSF drain (30 mL) with lumbar spinal puncture	1 day	Conservative treatment	Decreased level of consciousness	Dead
Schorl[14]	2009	Female	71	Meningioma	V-P shunt	Few days	Cranioplasty	Nonconvulsive status epilepticus, decreased level of consciousness (coma)	Recover
		Female	46	ACA infarction	V-P shunt	14 weeks	Cranioplasty	Hemiparesis, seizure	Recover
		Female	31	Traumatic subdural hematoma, postoperative infection	V-P shunt	1 day	Shunt valve adjustment, Shunt ligation, cranioplasty	Headache, hemiparesis	Recover
Jung et al.[9]	2012	Male	38	Traumatic epidural hematoma	CSF drain (20 mL) with lumbar spinal puncture	2 days	Conservative treatment	Decreased level of consciousness, hemiparesis	Recover

CSF: cerebrospinal fluid, SoT: Syndrome of the Trephined, V-P: ventriculoperitoneal, MCA: middle cerebral artery, ACA: anterior cerebral artery.

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Han et al.[7] reported the case of a 37-year-old man with traumatic brain injury and subsequent hydrocephalus who developed stupor after ventriculoperitoneal shunt surgery but recovered after cranioplasty. Jung et al.[9] described a case of a 38-year-old man with traumatic brain injury who developed a deep drowsy mentality and hemiparesis two days after lumbar drainage. Gschwind et al.[6] reported a 65-year-old man who developed mental deterioration after lumbar puncture and craniectomy for MCA infarction. Shen et al.[15] reported two cases of SoT after decompressive craniectomy and subsequent ventriculoperitoneal shunt placement for hydrocephalus.

There are some reports regarding SoT cases with lumbar spinal drainage, but in those cases, lumbar drainage was done at the moment of craniectomy, so the onset time or progression of SoT is not clear. Zhao et al.[17] found that 9 of 37 patients who underwent craniectomy and lumbar drainage developed SoT, and CSF volume may predict this condition. Motoyama et al.[12] compared 239 patients who received decompressive craniectomy with lumbar spinal drainage with patients who received decompressive craniectomy without lumbar spinal drainage. Downward brain herniation toward tentorial hiatus was more likely (10% vs. 3.3%) to happen in patients who received decompressive craniectomy with lumbar spinal drainage.

In this case, CSF flow diversion seemed to play a major role in the development and worsening of SoT. Initially, the patient experienced external hydrocephalus after a craniectomy. This condition occurs due to abnormal fluid collection caused by impaired CSF flow. Although external hydrocephalus occurs frequently, and most hydrocephalus cases resolve spontaneously, they can trigger external brain tamponade by adding mass effect upon the craniectomy site.[8,11] This mass effect, combined with atmospheric pressure, may compress the cortex underlying the skull defect and further interrupt CSF hydrodynamics.[1,16]

The patient underwent an L-P shunt operation, which was suspected to have caused CSF hypovolemia. Hygromas are also thought to cause intraventricular volume loss. A decrease in the CSF volume may result in a decrease in intracranial buoyancy, making the brain more susceptible to the downward forces of atmospheric pressure and gravity.[11,17] This, in turn, may lead to displacement of the brain, causing it to sink downward against hard intracranial structures such as the tentorium, falx, and foramen magnum.[6] If the patient loses more CSF fluid, the difference between the forces increases, probably resulting in paradoxical brain herniation, in which the brain becomes wedged between the hiatus of the tentorium.[15] This progression can be rapid and demonstrated by radiological examinations. When the niche of the tentorium is compressed, the CSF flow can be obstructed. If further CSF diversion from below occurs, from the level below the tentorium, such as in the L-P shunt in our case, this could form a negative pressure gradient across the tentorium, further worsening the herniation (**FIGURE 4**).

In the present case, we attempted to stop the CSF diversion using an L-P shunt, but this was not sufficient. Cranioplasty was required for recovery. This procedure was thought to reduce the pressure placed on the cortex, thereby decreasing the pressure gradient across the tentorium.[12]

CONCLUSION

Herein, we report an acute case of SoT after L-P shunt placement in a patient who had previously undergone decompressive craniectomy. The patient completely recovered after emergency cranioplasty but required a prolonged period of time.

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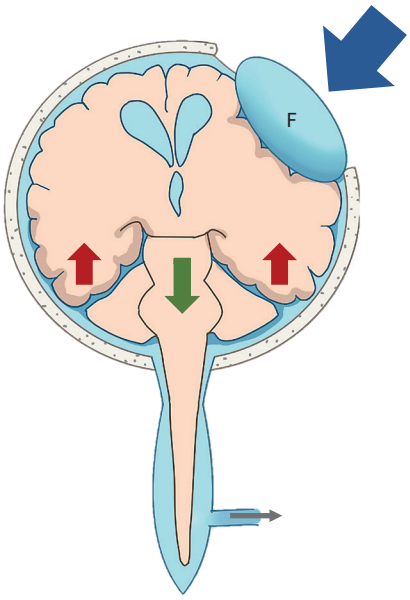

FIGURE 4. Schematic drawing of the possible mechanism of Syndrome of the Trephined (large blue arrow, atmospheric pressure compressing brain from the exterior; red arrows, buoyancy; green arrow, paradoxical cerebral herniation; small black arrow, cerebrospinal fluid leakage via the lumboperitoneal shunt).

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