ABSTRACT

Spontaneous intracranial hypotension (SIH) is usually a benign disease which mostly present as orthostatic headache and resolves by conservative treatment or epidural blood patch. However, in severe cases large subdural hematoma or brain caudal herniation can progress to brain herniation and neurologic complications. We introduce a rare case of SIH which presented as acute mental deterioration with unilateral acute subdural hematoma. A 60 years old female visited to emergency room for stuporous mental change and unilateral acute subdural hematoma. Decompressive craniectomy and hematoma removal was performed to release brain herniation and increased intracranial pressure. There was temporary improvement of consciousness, but sustained leakage of cerebrospinal fluid (CSF) and caudal brain herniation worsened patient’s condition. After recognizing that CSF leakage and hypovolemia was the underlying disease, emergent epidural blood patch and early cranioplasty was performed. After treatment CSF volume was normalized and patient recovered completely without neurologic deficits. Acute mental deterioration with unilateral subdural hematoma is a rare presentation for SIH. Treatment strategy for subdural hematoma with concomitant SIH patients, should be planned carefully with concerns to CSF hypovolemia and intracranial pressure.

Keywords: Intracranial hypotension; Cerebrospinal fluid leak; Subdural hematoma; Epidural blood patch

INTRODUCTION

Spontaneous intracranial hypotension (SIH) is a condition of cerebrospinal fluid (CSF) hypovolemia presenting as various symptoms. Dural loosening or defect and CSF-venous fistula at spine level causing CSF leakage is the etiology of this disease. Although the term is ‘intracranial hypotension’, the opening pressure of lumbar puncture is not always low (<6 cm H₂O) which demonstrates that low pressure is not the key mechanism of this disease. Most patients with SIH have benign course which is manageable with conservative treatment or epidural blood patch.
The spectrum of symptom is wide which makes the physician difficult to diagnose this disease. Orthostatic headache released by horizontal position is the cardinal symptom of SIH. Subdural hygroma can be developed due to compensation of low volume of CSF. Bridging vein tearing caused by caudal displacement of brain causes subdural hematoma. Brain sagging of the brain stem and cerebellum derived by loss of buoyancy causes cranial nerve palsy and mental deterioration.  

Brain magnetic resonance image (MRI) with gadolinium enhancement is effective diagnostic tool for SIH. Pachymeningeal enhancement, pituitary engorgement, venous engorgement of sinus, and subdural fluid collection are the key imaging signs of SIH. Caudal displacement of brain stem and cerebellum is best observed at the sagittal view. As SIH is necessarily caused by CSF leakage at spinal area, evaluation of CSF leakage site is important for treatment. Computed tomography (CT) myelography, heavily T2 weighted spinal magnetic resonance (MR) myelogram, radioisotope cisternography, and digital subtraction myelography has been introduced to detect the site of CSF leakage and underlying pathology.

Autologous epidural blood patch is known to be an effective treatment method by sealing the CSF leakage point and reserve CSF volume. Sometimes surgical dura reconstruction is needed in large CSF leakage site such as ventral dural tearing caused by osteophyte or disc herniation.

We document a rare case of SIH who visited to the emergency department with unilateral acute subdural hematoma causing acute mental deterioration. We wish this case could remind physician to keep in mind the possibility of SIH for patients presenting with unilateral subdural hematoma.

**CASE REPORT**

A 60 years old female visited to the emergency room for severe headache and vomiting followed by acute stuporous mental change. Onset of headache was 2 weeks prior aggravated by upright position and released by bed rest. Loss of consciousness occurred during evaluation at the emergency room. She had no major trauma history or concomitant disease. Non-contrast brain CT revealed a left convexity mixed type subdural hematoma. High and low density subdural fluid was mixed along the left side subdural space which indicates acute and chronic hematoma, causing right side midline shifting, approximately 12 mm (FIGURE 1A & B).

Emergent craniotomy and hematoma removal were performed immediately. Mixture of acute dark hematoma and hygroma-like fluid were observed at surgical field and completely evacuated. Brain surface was soft without significant edema or swelling. Post-operative CT revealed a complete evacuation of subdural hematoma and brain structure was well decompressed with correction of midline shifting (FIGURE 1C). Patient recovered its consciousness completely after the surgery. However, her consciousness gradually decreased and ended up unconscious 14 hours after the surgery. Emergent brain CT revealed a recurrent low density subdural fluid collection and small epidural hematoma with severe right side midline shifting (FIGURE 1D).

Emergent re-operation was performed by evacuating acute epidural hematoma and removing the bone flap for decompression (FIGURE 1E). Patient recovered its consciousness completely after the re-operation. However again, loss of consciousness occurred 12 hours after the operation.
re-operation. Contralateral side (right) oculomotor palsy has newly developed 3 days later and she remained unconscious for several days. Serial brain CT showed aggravation of brain sinking and effacement of basal cistern. According to serial brain CT and neurologic status, we suspected SIH at this point.

MR myelogram (heavily T2 weighted image) revealed a lateral extension of CSF at the left side of cervicothoracic junction level (FIGURE 2). Gadolinium enhance brain MRI revealed a diffuse dural enhancement, pituitary engorgement, venous sinus engorgement and effaced suprasellar cistern. Caudal displacement of brain stem and space narrowing of posterior fossa was clearly seen at the sagittal view of MRI. Effacement of suprasellar and pre-pontine cistern and decreased mamillo-pontine distance were detected (FIGURE 3). We finally diagnosed the patient for SIH caused by CSF leakage at cervicothoracic level. We acknowledged that skull

FIGURE 1. Time course of brain computed tomography. (A, B) Non-contrast brain computed tomography at the first visit to emergency department showing acute subdural hematoma mixed with low density fluid and multi-spectated layer. Mass effect of subdural hematoma affected severe midline structure shifting to the right side. (C) After the first surgery (craniotomy and hematoma removal), subdural hematoma was evacuated and the midline structure was normalized. (D) Fourteen hours after the first surgery, the brain CT revealed a low density subdural fluid collection and multiple small epidural hematoma. (E) Emergent decompressive craniectomy was performed, and immediate post-operative CT showed a release of brain compression and midline shifting. (F) Two weeks after the second surgery, left hemisphere compressed to the right side and midline shifting was worsened. CT: computed tomography.
defect state has aggravated brain sinking and planned for early cranioplasty. Autologous epidural blood patch at cervicothoracic junction (12 mL) and cranioplasty was performed simultaneously. Patient was placed in the Trendelenburg position to increase intracranial CSF volume. After treatment, patient neurologic status improved gradually and eventually recovered completely without neurologic deficit. One year follow up brain MRI showed complete resolution of all findings that indicated SIH (FIGURE 3).

**DISCUSSION**

We introduced a rare case of SIH presenting as acute mental deterioration and unilateral acute subdural hematoma. Decompressive craniectomy has worsened caudal displacement of brain structure by exposing to atmospheric pressure. Fortunately, targeted autologous epidural blood patch and early cranioplasty has normalized intracranial CSF volume and patient recovered completely without any neurologic deficits. However misdiagnosis of SIH led to inappropriate treatment and worsened patient’s condition.

Spectrum of subdural fluid collection is various in SIH patients.\(^3,^6,^{15}\) Subdural hygroma is derived from extravasation of fluids as a result of compensation for CSF hypovolemia. Most of the patients present as ‘bilateral’ lesion because CSF hypovolemia is associated with both hemisphere. Bridging vein tearing can be caused by caudal displacement of brain parenchyma and usually present as chronic or subacute hematoma. Among patients with chronic subdural hematoma caused by SIH, bilateral lesion was reported to be approximately 90\%.\(^6,^{15}\) We missed to suspect SIH because of both ‘unilateral’ and ‘acute’ subdural hematoma presentation. Furthermore, acute mental deterioration is uncommon for SIH because it usually presents as chronic subdural hematoma with gradual symptom progress.
Osada et al.\textsuperscript{14} reported a case series of 4 patients whom developed coma and transtentorial herniation with unilateral chronic subdural hematoma due to SIH. Unilateral subdural hematoma was explained by the asymmetrical cranium and the downward pulling force caused by CSF hypovolemia. Among 4 patients, Duret hemorrhage and posterior cerebral artery territory infarction occurred as a result of brain stem compression and herniation. Common features of these patients were young age, unilateral lesion, no trauma history, and acute mental deterioration with severe brain herniation. Patient’s condition did not improve after surgical evacuation and recollection of subdural fluid or hematoma occurred immediately which is similar to current case.\textsuperscript{14} Acute mental deterioration is an alarm sign of brain stem compression and herniation. In this condition accompanied with SIH, the treatment should be urgent to avoid permanent neurologic deficits associated with brain herniation.

In our case, decompressive craniectomy worsened brain sinking which mimics with the condition called ‘sinking brain syndrome’.\textsuperscript{7} According to the Monro-Kellie hypothesis, when the brain is enclosed by the skull bone, the sum of intracranial structure such as brain parenchyma, arterial/venous structure, and CSF volume remains constant.\textsuperscript{13} Vascular hyperemia, especially venous engorgement occurs for compensation of CSF hypovolemia. However, when skull bone is removed, Monro-Kellie hypothesis is no longer applied because the atmospheric pressure is involved. Exposure to atmospheric pressure and CSF hypovolemia state will increase the negative intracranial pressure and exacerbate caudal brain herniation. We performed an early cranioplasty to protect the brain from exposed atmospheric pressure after we recognize that CSF leakage and hypovolemia was the
underlying problem. CSF hypovolemia should be suspected in patients with early brain sinking after craniotomy or craniectomy surgery. In several case reports, severe caudal herniation of brain stem and cerebellum occurred after evacuation of subdural fluid collection which led to permanent neurologic deficits or death. Misdiagnosis of SIH and inappropriate surgical treatment exposing to atmospheric pressure may lead to life-threatening condition by worsening caudal herniation.

Interesting finding in our case was that the patient’s consciousness completely recovered right after evacuation of subdural hematoma. However, recovery was temporary and did not last for 12 hours after both surgeries. Temporary recovery of consciousness after surgery represents that although the CSF volume was low, the intracranial pressure was high due to the mass effect of subdural hematoma. It seems that removal of subdural hematoma released the increased intracranial pressure temporarily, but underlying CSF hypovolemia induced caudal herniation re-worsened its condition. Sealing the CSF leakage point without surgical evacuation is more safe and effective in patients with normal or low intracranial pressure. However, if the intracranial pressure is high, subdural hematoma should be surgically evacuated to release brain compression. The dilemma is that estimating the intracranial pressure by brain image is limited and measuring lumbar opening pressure contains the risk for caudal herniation of brain structure, especially for increased intracranial pressure state. Trendelenburg position may improve neurologic deficits in condition of low intracranial pressure state with CSF hypovolemia. Loya et al. suggested that failure of symptom improvement after Trendelenburg position with large subdural hematoma should be evacuated emergently. If surgical evacuation is necessary, epidural blood patch should be performed prior to surgery, and Trendelenburg position must be maintained after.

Current case showed dramatic change of brain MRI after normalization of CSF volume (FIGURE 3). Absent of blood brain barrier at pachymeninges is involved to extravasation of contrast material which is the action of compensation for CSF hypovolemia. In the other hand, leptomeninges is not involved because blood brain barrier exists. Furthermore, engorgement of venous sinus and enlarged pituitary gland are important sign of vascular compensation to CSF hypovolemia. In severe CSF hypovolemia state, the loss of CSF buoyancy leads to downward sagging of brain stem and cerebellum. Downward force derived by loss of CSF buoyancy leads to mammillary body descents and narrow the space between mammillary body and superior border of pons (mamillo-pontine distance). Effacement of suprasellar or pre-pontine cistern is also commonly detected. Decrease of the angle between vein of Galen and straight sinus is a hallmark of posterior fossa crowding. Sometimes cerebellar tonsill descent to the cervical spinal canal which could be confused as Chiari malformation. For SIH patients, brain MRI with gadolinium enhancement is effective for diagnosis and also for evaluating the treatment response.

In our case, heavily T2 weighted MR myelogram successfully detected CSF leakage point at cervicothoracic junction. MR myelogram has an advantage of non-invasiveness compared to other diagnostic tools. In condition of increased intracranial pressure, lumbar puncture can develop a serious caudal displacement and herniation. Kranz et al reported that only 34% of patients were measured for low CSF pressure (<6 cm H2O) in SIH patients. Measuring intracranial pressure does not help significantly in diagnosis and may increase the likelihood of complication. The possibility of caudal herniation after lumbar puncture should be considered, especially for subdural hematoma patients.
Clinical suspicion of SIH is the most important step for physician to deal with this disease. Important clues for SIH are young age, no major trauma history, history of positional headache, posterior neck pain and stiffness, and cranial nerve palsy. Therefore, a detailed interview of past symptoms and neurologic examination are important to suspect this disease. In condition of subdural fluid collection accompanied with SIH, estimating the intracranial pressure is very important to decide surgical option. Surgical evacuation should be performed if the intracranial pressure is high, but if it is low, surgical treatment can worsen CSF hypovolemia and caudal displacement of brain structure.

CONCLUSION

Acute mental deterioration with unilateral subdural hematoma is a rare presentation for SIH. Treatment strategy for subdural hematoma with concomitant SIH patients, should be planned carefully with concerns to CSF hypovolemia and intracranial pressure.

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