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CASE REPORT

Spontaneous intracranial hypotension complicated by unilateral subdural hematoma, coma, and the rare Kernohan's notch phenomenon

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Abstract

Spontaneous intracranial hypotension (SIH) is a highly underdiagnosed condition with a wide range of manifestations. Although SIH has traditionally been viewed as having a benign disease course, recent evidence suggests otherwise. In this case report, we present a 71-year-old woman who was previously treated with a tapered dose of corticosteroids for an episode of SIH with behavioral manifestations. In this current report, we present her second episode which was complicated by a unilateral sub-acute subdural hematoma leading to loss of consciousness and coma. While the patient regained full consciousness and recovered considerably, she still suffers from ipsilateral hemiparesis as a result of Kernohan's notch phenomenon 2 years post-hospitalization. To the author's knowledge, this is the first case to involve persistent motor deficit after the resolution of SIH. We believe that surgical hematoma evacuation prior to addressing the cerebrospinal fluid leak may have contributed to aggregated downward displacement of the brain precipitating Kernohan's notch phenomenon.

KEYWORDS

coma, epidural blood patch, Kernohan's notch phenomenon, spontaneous intracranial hypotension, stroke, subdural hematoma

1 | BACKGROUND

Spontaneous intracranial hypotension (SIH) is often characterized by the sudden onset of headaches with postural worsening when standing from a recumbent position.[1] Headaches caused by SIH are attributed to engorgement of the pain-sensitive cerebral venous sinuses, which is a possible pathophysiological response to the loss of cerebrospinal fluid (CSF). This response is best explained by the Monro-Kellie doctrine of constant intracranial volume between the three components: The brain, CSF, and intracranial blood. When there is a loss of CSF volume, it triggers a compensatory increase in the volume of either one of the other two compartments, or both in an attempt to keep the intracranial volume stable.[2]

SIH has traditionally been perceived to be a benign and self-resolving condition. However, within the past decade, our improved understanding of its complexity and atypical clinical manifestations have demonstrated that SIH has been a highly underdiagnosed and underestimated

clinical entity. Besides postural headaches, the condition has been associated with nausea, emesis, visual and ocular problems, taste alterations, sleep attacks, chorea, behavioral changes, dementia, fatigue, and even coma.[1,3–5] The classic diagnostic signs include a triad of postural headaches, low opening CSF pressure, and cranial magnetic resonance imaging (MRI) showing diffuse pachymeningeal enhancement[4] and possibly herniation of the cerebellar tonsils, creating the appearance of a 'brain sag'.[3,5] While the cerebrospinal fluid leak is believed to be the cause of SIH, a leak is not always identified in spinal imaging, creating a diagnostic challenge. Furthermore, there are an increasing number of reports showing normal opening pressures during a spinal tap, challenging one of the hallmark features of the condition.[3,5,6] Diagnostic imaging can also be misleading due to patient positioning during cranial MRI scans. A cranial MRI scan performed when the patient is placed supine can possibly mask or underestimate the downward displacement of brain structures, which can provide a possible explanation as to why herniation of the cerebellar tonsils is not evident on MRI studies in a considerable number of patients suffering from SIH.[7]

A downward displacement of the brain structures because of SIH can potentially cause the bridging veins to stretch and rupture, resulting in bleeding and the formation of a subdural hematoma (SDH).[6] Recent literature on SIH has increasingly reported this as a possible complication of the condition. Perhaps this is not due to its recent increased occurrence, but rather due to our increased awareness of the atypical manifestations of the clinical entity. While management of SDH is usually a clinical decision based on its etiology (traumatic or non-traumatic), size of the hematoma, the patient's general and neurological condition, and whether any cerebral herniation is present,[8] its occurrence in the context of SIH can further complicate this decision. A few case reports, including the current one, are reporting clinical deterioration of patients after hematoma evacuation in the background of SIH as a result of aggravated downward displacement of the brain structures.[9–11] Therefore, SIH should be kept as a differential diagnosis when patients present with SDH and a history of headaches, especially when accompanied by atypical neurological presentations. This can help avoid a surgical intervention which can further complicate the patient's clinical condition.

2 | CASE PRESENTATION

A woman in her 70s, who was previously treated for spontaneous intracranial hypotension with a tapered course of corticosteroids and full recovery,[3] had an abrupt onset of thunderclap headaches coupled with elevated blood pressure. On the third day of her headaches, the patient complained of nausea and diarrhea which prompted a visit to the bathroom, and within 10 min, she was found unresponsive on the floor by her husband. There was no history of trauma.

The patient's previous episode of SIH in 2017 presented with headaches, sleep attacks, chorea, urinary incontinence, and dementia-like behavioral manifestations.[3] Due to her atypical presentation, the diagnosis was delayed by more than a year. Within this period, frontotemporal dementia and narcolepsy were considered likely diagnoses by a number of neurologists. However, the full range of symptoms could not be explained by either condition alone. The patient's past medical history was insignificant except for a cholecystectomy in 1992 and a radical hysterectomy in 1997 due to multiple fibroids. She gave birth to three children via natural deliveries. She is a non-disease carrier of the beta thalassemia trait. Her family history included the myocardial infarction of her mother at the age of 76 and a pacemaker insertion of her brother in his 70s. However, there was no personal or family history of neurological or neurovascular conditions.

When the patient presented to us in 2017, we performed a series of tests including a lumbar puncture with a normal opening pressure of 10 cmH_2O, cerebrospinal fluid (CSF) analysis without any remarkable findings, and an extended extractable nuclear antigen panel with all negative results. It was after a re-assessment of her previous cranial MRI studies from her previous doctor visits that we were able to reach a diagnosis of SIH. We offered a tapered course of corticosteroids as an initial treatment. While the literature does not provide a clear role for corticosteroids in the management of SIH, the medical approach was agreed upon with the patient and her family as a non-invasive initial approach. The patient's recovery was gradual with normalizing bladder control within a month and the disappearance of chorea and behavioral issues around 8 weeks. She was on 80 mg prednisolone with a 10% reduction in dose every other week. The patient fully recovered and the medical treatment was ended after a period of 4 months.

While the current episode was similar in the way that headaches manifested, the rest of her previous symptoms were absent. As a result, the patient assumed her headaches were a result of hypertension and used antihypertensive medication at home without seeking medical attention. Upon admission, her Glasgow Coma Score (GCS) was determined to be 4/15 (E1V1M2). Her blood pressure was 145/95 mmHg, oxygen saturation was 94% in room air and temperature was 36.4°C. Her breathing pattern was described as apneustic. The patient was unresponsive to any stimulus. An emergency computed

tomography (CT) scan of the cranium was organized, which showed a large unilateral right-sided sub-acute subdural hematoma with a marked right-to-left midline shift (Figure 1A). An emergency craniotomy was performed and sufficient brain expansion was reported after hematoma evacuation. Postoperatively, the patient remained unresponsive yet hemodynamically stable over the next few days. A follow-up cranial CT scan 48 h after the surgery showed a larger hematoma and no improvement of the midline shift (Figure 1C). A reassessment of the previous cranial CT imaging indicated a missed diagnosis of cerebellar tonsillar herniation as evidenced by the entire foramen magnum being filled with soft tissue density and the lack of hypodense cerebrospinal fluid (CSF) (Figure 1B). Following this discovery, a spinal MRI was ordered to detect a possible CSF leak site. A cranial CT was also ordered for follow-up. On imaging, foramen magnum appeared to have a lower soft tissue density (Figure 1E) and both the volume of hematoma and the midline shift appear to have reduced (Figure 1D). Unfortunately, the spinal MRI study was inconclusive, and a dynamic CT scan was not available. Given the likely diagnosis, it was decided among the medical team that placing the patient in the Trendelenburg position could induce a rapid increase in intracranial pressure[12] and help reduce the effects of downward displacement.

Identifying the leak would allow for a possible targeted epidural blood patch to seal the leak and reverse the

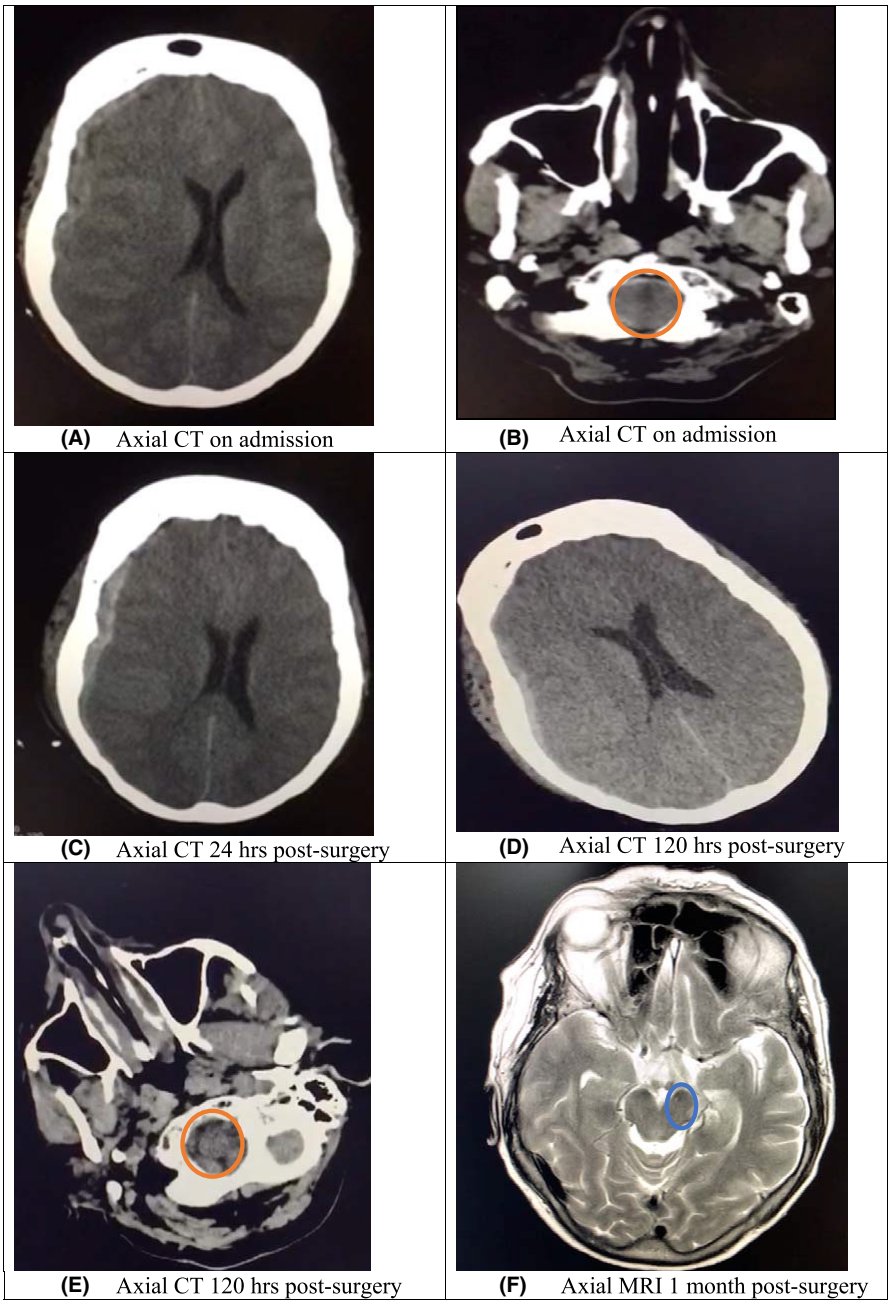
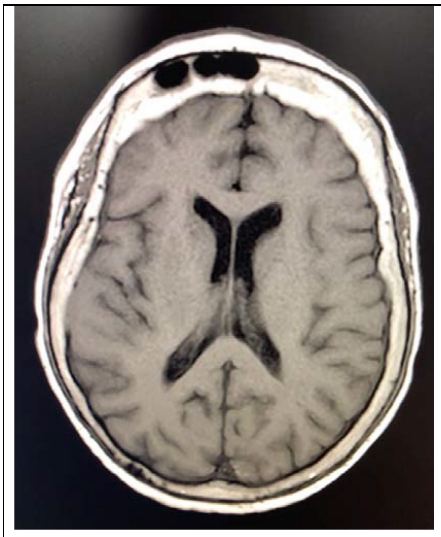
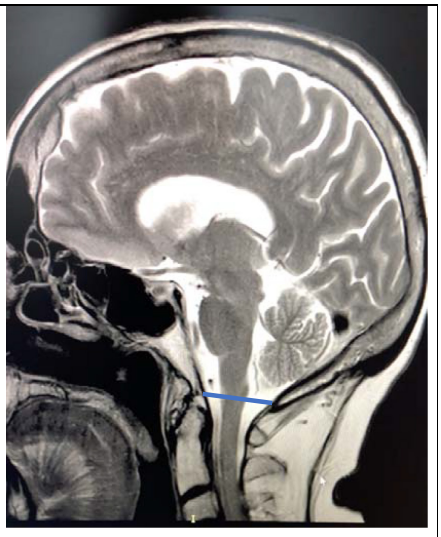


FIGURE 1 Non-contrast cranial CT imaging prior to and post hematoma evacuation. (A) This axial non-contrast CT brain scan shows a sub-acute, right-sided subdural hematoma with mass effect at the time of admission. (B) This axial non-contrast CT brain scan shows increased soft tissue density in the foramen magnum and the lack of hypodense cerebrospinal fluid at the time of admission, marked by the circle. (C) This axial non-contrast CT brain scan shows a sizeable acute right subdural hematoma, with moderately severe mid-line shift, 2 days after emergency craniotomy. (D) This axial non-contrast CT brain scan shows a reduced sub-acute right subdural hematoma, with moderate mid-line shift, 5 days after emergency craniotomy. (E) This axial non-contrast CT brain scan shows a relatively reduced soft tissue density in the foramen magnum with the appearance of some hypodense cerebrospinal fluid, 5 days post emergency craniotomy, marked by the circle. (F) This axial non-contrast CT brain scan shows a lesion on the left cerebral peduncle (marked by the circle), which appears to be responsible for the false localizing sign.



(A) Axial MRI Imaging



(B) Sagittal MRI Imaging

FIGURE 2 Axial and sagittal MRI imaging at 1-year. (A) This axial non-contrast MRI brain scan shows complete resolution of the right subdural hematoma and no mass effect. (B) This sagittal MRI brain scan shows normal positioning of the cerebellar tonsils and no evidence of herniation into the foramen magnum.

condition.[13] Instead, a blind lumbar epidural blood patch was performed at the L3-L4 level with 20 ml autologous blood. The patient was kept in the Trendelenburg position to aid with the upstream movement of blood in the epidural space to help seal a possible cerebrospinal fluid leak.[1] Within a few hours, the patient started responding to painful stimuli. Her GCS improved to 6/15 (E2V1M3). After 48 hours, a second untargeted lumbar blood patch was performed with a longer period in the Trendelenburg position. Approximately 8 h after the procedure, the patient started opening her eyes when prompted with speech and started making incomprehensible sounds. Her GCS was measured as 8/15 (E3V2M3). The patient was kept in the Trendelenburg position for a total of 48 hours. At the end of this period, her GCS had improved to 12/15 (E4V4M4). Over the course of the next few days, she moved up to 15/15 on GCS. However, the patient exhibited symptoms of dysarthria and dysphagia and remained ipsilaterally hemiplegic, suggesting compression of the contralateral cerebral peduncle against the cerebellar tentorium at a level above the level of decussation, causing an indentation, also known as the Kernohan's notch phenomenon.[14] This was later demonstrated by MRI imaging at 1-month follow-up (Figure 1F). Before discharge, a third epidural blood patch was planned at a higher spinal position (T7–T8) to reach a possible leak that could be at the cervical level. Nevertheless, this attempt also did not produce any immediate results. The patient showed slow but gradual improvement in her motor function over the course of the following months.

After discharge, the patient was taken home after necessary arrangements had been made for her care. She would be kept in Trendelenburg position at night for a total of 8 h for the following few weeks. The patient showed marked improvement in her mental status and speech over the next weeks. Hemiplegia resolution was quite slow, unlike the reports in literature.[4–6,9–11] At the 1-year mark, cranial MRI has remained negative for any SIH-related pathologies (Figure 2). While the patient has no other apparent deficits, she remains hemiparetic with marked weakness in her right limbs despite an aggressive physiotherapy regimen. However, she is mobile with a Zimmer frame and can attend to her own personal care.

3 | DISCUSSION

Despite the growing volume of literature on SIH, the condition still provides a diagnostic challenge due to its highly variable clinical presentation. As a consequence, a standard management strategy does not exist. Instead, an individualized approach is used based on each patient's clinical presentation and disease severity.[15] In a certain group of patients, successful management via bed rest, hydration, and NSAIDs is possible when the headache is the only presentation.[4] The use of oral or intravenous caffeine and theophylline has shown mixed results with some possible benefits but not enough to make them a suitable first-line treatment modality.[16] The use of corticosteroids for the management of SIH has also been somewhat controversial. However, there is a growing number of publications, including our previous case report, reporting the complete recovery of patients with headaches accompanied by neurological manifestations after a course of corticosteroids.[3,6,17–22] Three mechanisms have been described for the role of corticosteroids in the management of SIH. As an anti-inflammatory medication, corticosteroids can help reduce cerebral edema and

inflammation caused by the downward displacement of the brain.[18] Second, corticosteroids are known to induce fluid retention. This can potentially have a role in reducing meningeal inflammation and vascular leakage.[19] Finally, corticosteroids are believed to help reabsorb CSF from extradural space, helping increase CSF volume.[20]

While conservative and medical therapies exist, epidural blood patching is often accepted to be the mainstream treatment for SIH. A large number of cases either use epidural blood patches as first-line management or secondary alternative when conservative management or medical therapy does not help.[23] Epidural blood patches have been reported to lead to an immediate resolution of clinical signs and symptoms with success rates as high as 83%.[23,24] Nevertheless, in our case, multiple blood patches which were targeted at different spinal positions did not yield any immediate benefits. Rather, we observed that Trendelenburg position was of substantial benefit in recovery. The longer we kept the patient in this position, the more improvements we observed during her hospitalization.

Our patient was rushed to the emergency room because of loss of consciousness and an immediate decision was made by the neurosurgeon to evacuate the SDH as it was associated with a significant shift of the midline cerebral structures. However, the increased size of hematoma and the patient remaining unconscious postoperatively prompted a reassessment and that is when we were consulted. Literature provides a few case reports of patient deterioration after hematoma evacuation because of SIH.[11,25,26] This may be explained by the aggravated downward herniation of the brain once the hematoma was removed, which further exacerbated the already low intracranial pressure.

Cases reported in the literature have all been described to recover almost immediately once they received an epidural blood patch. We believe that our case stands out by the fact that aggravated downward displacement of the brain contributed to compression of the contralateral crus cerebri in the midbrain by the free edge of the herniating tentorium.[27] This is an uncommon occurrence known as the Kernohan's notch phenomenon. This phenomenon explains the ipsilateral hemiparesis as a false localizing sign. The persistence of the patient's loss of motor function is believed to be the result of permanent damage to the motor fibers because of this phenomenon.

AUTHOR CONTRIBUTION

AO has written the paper in its entirety.

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DATA AVAILABILITY STATEMENT

All the laboratory data and imaging studies are available per request.

CONSENT

Patient's permission (both oral and written) has been obtained for the publication of relevant medical data by protecting anonymity.

ETHICS STATEMENT

The author declares no conflict of interest. This article is the original work of the author and is not being considered for publication in any other journal. The paper reflects the author's own research in a truthful manner. All sources have been properly disclosed and given credit to.

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