CASE REPORT

Deep venous structures distortion in spontaneous intracranial hypotension as an explanation for altered level of consciousness

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SUMMARY

Spontaneous intracranial hypotension (SIH) is a syndrome of low pressure headache associated with low CSF pressure. The condition is generally considered benign but extreme cases of SIH can lead to changes in the level of consciousness. We describe a case in which alteration in the level of consciousness was prolonged and severe, and could not be explained solely by the presence of subdural collections. MRI of the brain showed evidence of impaired venous flow secondary to brain sagging causing distortion of deep venous structures.

BACKGROUND

Spontaneous intracranial hypotension (SIH) is the syndrome of low pressure headache associated with low CSF pressure in the absence of dural puncture. The syndrome is characterized by severe headache that is postural in nature, provoked by sitting or standing, and relieved by a recumbent position. Several associated symptoms such as postural tinnitus, vertigo, nausea, mild stiffness, and radicular pain have been reported.¹ The condition is generally considered benign but extreme cases of SIH can lead to changes in the level of consciousness.² The conventional treatment options for SIH are bed rest and blood patch. Surgery is reserved for patients with persistent leak despite blood patch and those with alteration of level of consciousness secondary to significant subdural collections.³ We describe a case in which alteration in the level of consciousness was prolonged and severe, and could not be explained solely by the presence of subdural collections.

CASE PRESENTATION

A 31-year-old previously healthy man was seen by the neurology service for recent onset headache. The headache started 3 weeks prior to presentation. The patient had a global headache that was initially mild to moderate in intensity. It progressed to a severe bifrontal headache and was associated with nausea but no vomiting. The headache was worsened by walking, climbing stairs, coughing, or sneezing. Lying flat and the use of mild analgesics relieved the headache symptoms. The patient had no significant past medical history and no history of trauma.

Initial laboratory investigations, including complete blood count, and renal and coagulation profiles were unremarkable.

MRI of the brain (figure 1A) showed a small extra-axial fluid collection. It also demonstrated smooth dural enhancement and sagging of the brain and posterior fossa structures, suggestive of low intracranial pressure (ICP).^{4–6} The patient was admitted for workup, and a lumbar puncture was performed yielding bloody CSF. The opening pressure was 8 cm H₂O with normal glucose and protein.

The patient continued to have worsening headache and started to be confused and drowsy. CT of the head was obtained (figure 1B), showing bilateral subdural hematomas with interval increase in size on the left side causing a significant mass effect, midline shift towards the right, and effacement of the basal cisterns. Because of the deteriorating neurological status and the mass effect, the patient underwent an urgent left frontotemporo-parietal craniotomy. A large subdural fresh clot was found mixed with dark chronic hematoma fluid. Postoperatively, the patient was awake and oriented with no focal or lateralizing deficits. A post-operative CT scan was obtained showing good evacuation of the hematoma.

On the second postoperative day, the patient was found to be unresponsive. An urgent CT scan was done showing a compressive extra-axial collection with significant midline shift. Reopening of the craniotomy was done showing a fresh clot, which was evacuated, and the brain was found not to be under pressure. An ICP monitor was inserted. The patient was fully awake after the second craniotomy with no focal neurological deficits, and ICP was approximately 10 cm H₂O. Over the next 3 days the patient continued to experience a decrease in his level of consciousness. Repeated CT scans were however stable and failed to demonstrate significant subdural mass lesion. The patient underwent a blood patch of 23 mL that was injected between thoracic levels 11 and 12. The patient showed little improvement in fluctuations of his level of consciousness. A complete spinal MRI and nuclear medicine cisternography were done which did not reveal a source for the CSF leak.

On the second day after the blood patch, the patient was placed in a Trendelenburg position (–15°). In this position, the patient had a sustained normal level of consciousness. The patient was kept in the Trendelenburg position for 48 h, after which he was gradually elevated to a flat position, then 15–30° tilt, 30–45° tilt, then sitting, and then mobilizing, over a period of 72 h.

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Figure 1 Axial enhanced T1 MRI, before admission, demonstrated bilateral dural enhancement and small extra-axial collection (A). These findings are suggestive of low intracranial pressure. Preoperative plane CT scan, during acute deterioration, showed a significant increase in the left side hematoma with an acute component with a significant midline shift and effacement of the basal cisterns (B). T2 weighted MRI images during the acute phase of neurological deterioration showed brain sagging and brain edema involving the corpus callosum (arrow) (C). Axial proton density sequence showed edema involving in the corpus callosum (arrow) (D) and the midbrain (arrowhead) (E). MR venography showed the straight sinus being significantly displaced upward to an almost vertical orientation. The internal cerebral veins were displaced downward, creating a kink between them and the vein of Galen, which could not be well visualized (F).

MRI and MR venography evaluations were obtained during and after the acute phase of neurological deterioration. The initial MRI showed bilateral holohemispheric subdural

collections, downward herniation of the cerebellar tonsils, crowded foramen magnum, effacement of the basal cisterns, brain sagging, and effacement of the suprasellar cistern (figure

1C). It also showed a focal area of T2 hypersignal involving the splenium of the corpus callosum (figure 1C, D). There was also an increased signal on the proton density sequence of the left superior colliculus and the left lateral aspect of the pons and mesencephalon (figure 1E). MR venography showed the straight sinus being significantly displaced upwards to an almost vertical orientation. The internal cerebral veins were displaced downward, creating a kink between them and the vein of Galen, which could not be well visualized (figure 1F). Repeat MRI 16 days later showed stable size of the subdural collection but improvement in the cerebellar tonsil herniation and brain sagging, resulting in decreased crowding of the foramen magnum and resolution of the effacement of the basal cisterns (see online supplementary figure 2A). There was also interval resolution of the T2 signal abnormality involving the brainstem and the splenium of the corpus callosum (see online supplementary figure 2B, C). Kinking of the vein of Galen was still present but this vein was now well visualized (see online supplementary figure 2D). The patient was discharged home. At his 6-month follow-up evaluation, he showed no neurological deficits and no residual headache.

DISCUSSION

SIH was first described in 1938 by Schaltenbrand.⁷ It occurs in 2–5 per 100 000 of the population, with a male to female ratio of 1.5–2:1. The most common age of presentation is between 30 and 50 years of age. It has no clear etiology. This has resulted in numerous terms in the literature, such as spontaneous low CSF pressure, spontaneous CSF hypovolemia, spontaneous or essential aliquorrhea, spontaneous CSF hypovolemia, low CSF pressure headache, spontaneous spinal CSF leak, and intracranial hypotension headache.^{2 7–11} The basic pathophysiological premise of SIH is the presence of a spontaneous CSF leak. The precise cause of the CSF leak remains largely unknown, but an underlying structural weakness of the spinal meninges is generally suspected.¹

The workup for SIH is aimed initially at making an accurate diagnosis and secondly in identifying the source of the CSF leak. Standard radiological workup, using CT and MRI scans, shows a combination of the following features: subdural fluid collections, enhancement and thickening of the pachymeninges, engorgement of venous structures, pituitary hyperemia, and sagging of the brain. The specific workup for identification of the site of the leak includes spinal MRI, CT and/or MRI myelography/cisternography, and radionuclide nuclear medicine cisternography.

In SIH patients, the lumbar puncture opening pressure can be <6 cm H₂O, which is low by definition. However, the majority of patients with SIH have a normal CSF opening pressure. The Trendelenburg position can be a useful screening tool for SIH cases presenting with headaches.¹²

Treatment of SIH depends, for the most part, on the clinical presentation of the patient. Conservative measures used in the treatment of SIH include bed rest, oral hydration, and caffeine and pain medication to control the headache.

In the case of persistent symptoms, epidural blood patch or surgical repair of the leak site is recommended. Acute deterioration of the level of consciousness, although considered rare, represents a challenging scenario, necessitating more aggressive measures, with possible craniotomy and evacuation of the hematoma or collection in order to relieve the cause of the new rise in ICP. Intrathecal infusion of saline has been described as a therapeutic strategy to reverse the CSF volume loss and correct the hydrodynamic derangement.¹³

Our patient represents an extreme case of SIH, with deterioration of his level of consciousness, persistent even after a blood

patch and evacuation of the subdural hematoma. Indeed, despite a first attempt at treating the suspected CSF leak with a blood patch and correction of the mass effect with evacuation of the subdural hematoma, our patient remained drowsy and at times stuporous. We postulated that the kinking of the vein of Galen seen on MRI could be causing a blockage or impaired flow through the vein, leading to deep cerebral venous hypertension with secondary edematous changes in the splenium of the corpus callosum, left superior colliculus, and the left lateral aspect of the pons and mesencephalon. These changes in turn could explain the observed decreased level of consciousness in our patient. In the hope of correcting the venous distortion, we opted to place our patient in the Trendelenburg position. This led to a spectacular improvement in our patient's level of consciousness. Other causes of a decrease in consciousness in some SIH patients are brainstem compression caused by brain sagging and the mass effect caused by large extra-axial collections.¹⁴

Two other case reports mentioned the Trendelenburg position for refractory SIH.^{15 16} Ferrante *et al* reported the application of a lumbar epidural blood patch and Trendelenburg position simultaneously in an SIH obtunded patient. They speculated that this may facilitate the spread of the blood patch into the epidural space.¹⁷ Our case is the first to demonstrate improvement in the clinical picture and resolution of the brainstem edema in the acute phase of illness in refractory SIH by placing the patient in the Trendelenburg position. This could be attributed to the stabilizing effect of the Trendelenburg position on CSF pressure at the leak site by re-establishing the intracranial–intraspinous pressure gradient of CSF pressure. It further supports the theory that a decreased level of consciousness can be caused by brain sagging, venous distortion and obstruction, and associated swelling of the brainstem structures, rather than being secondary to increased ICP caused by the subdural collections. The Trendelenburg position can be added in the armamentarium of treatment options for refractory cases of low ICP associated with alteration in the level of consciousness.

Learning points

- ▶ Spontaneous intracranial hypotension (SIH) is an increasingly recognized entity, especially in the past decade.
- ▶ SIH is often under diagnosed, and the diagnosis may be significantly delayed.
- ▶ Decreased level of consciousness is an uncommon clinical presentation, not always caused by the mass effect from a subdural collection, but rather by impaired deep cerebral venous flow secondary to brain sagging which can lead to venous hypertension.
- ▶ The Trendelenburg position can be used in refractory cases as an adjunct treatment option in order to accelerate the recovery of the patient with a potentially dangerous altered level of consciousness.

Contributors All authors contributed to this manuscript. AMA: conception, design, acquisition of the data, analysis of the data, and drafting the article. HA-J: conception, design, and drafting the article. CT: interpretation of the data, and revising the paper critically for important intellectual content. JM: conception and design, interpretation of the data, and drafting the article.

Competing interests None.

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