Spontaneous intracranial hypotension: improving recognition and treatment strategies in the local setting

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A B S T R A C T

We report a case of spontaneous intracranial hypotension with classic symptoms of orthostatic headache and acute presentation of subdural haematoma on computed tomographic scan. Conventional approach with conservative treatment was initially adopted. The patient's condition, however, deteriorated after 2 weeks, requiring surgical evacuation of the intracranial haemorrhage. We reviewed the clinical features of this disease and the correlated magnetic resonance imaging findings with the pathophysiological mechanisms, and described treatment strategies in the local setting. Subtle findings on initial computed tomographic scan are also reported which might improve pathology recognition. Spontaneous intracranial hypotension is not uncommonly encountered in Hong Kong, and physicians must adopt a high level of clinical suspicion to facilitate early diagnosis and appropriate management. In addition, novel therapeutic approaches may be required in those with recurrent symptoms or who are refractory to current treatment strategies.

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Case report

A 54-year-old man with no history of trauma was admitted to the Prince of Wales Hospital for headache of progressive severity accompanied by dizziness in August 2012. He had consulted the emergency room 4 weeks earlier for neck pain, and had an unremarkable computed tomographic (CT) scan of the brain (CTB). Further enquiry revealed an orthostatic component within the headache (worse in upright position and relieved within minutes of assuming supine posture), while admission CTB revealed interval development of bilateral 5 mm–thick frontoparietal subacute subdural haematomas (SDHs) with disproportionate tightness of the basal cisterns.

Cerebral magnetic resonance imaging (MRI) additionally demonstrated compression of the midbrain, but no caudal herniation of the cerebellar tonsils beyond the foramen magnum. Contrast study showed diffuse pachymeningeal enhancement, venous sinus distension, and prominent pituitary gland. Spinal MRI was unremarkable and MRI cisternography/myelography was negative for cerebrospinal fluid (CSF) leakage.

The patient was advised complete bed rest with adequate hydration. His neurological status was intact all along. However, he reported persistent, severe bifrontal headache which, after 2 weeks, was accompanied with repeated projectile vomiting. Computed tomographic scan of the brain at this juncture revealed enlargement of the SDH with development of acute haemorrhage. Emergency evacuation of subdural blood was performed with development of low intracranial pressure during the evacuation process.

The patient's symptoms improved markedly thereafter, and CTB reassessment showed minor residual blood. The patient was discharged in a neurosurgically stable condition, and currently remains asymptomatic.

Discussion

Intracranial hypotension is traditionally attributed to leakage of CSF from a dural defect along the craniospinal axis, which can occur spontaneously, such as due to rupture of Tarlov cyst[1] or dural weakness in connective tissue disorder.[2] Intracranial hypotension can also be precipitated by direct trauma or iatrogenic causes such as a lumbar puncture. The commonest cause, however, is a spontaneous defect

自發性顱內低壓：提高識別和改善治療策略

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本文報告一宗自發性顱內低壓的病例，病人出現姿勢性頭痛和電腦斷層掃描顯示急性硬膜下血腫的典型症狀。起初使用保守治療的傳統方法，可是兩週後患者的病情惡化，須進行開顱手術把血腫清除。本文回顧這種病的臨床特點和與病理生理機制的相關性磁共振成像結果，並描述本地的治療策略；並會提出在最初的電腦斷層掃描上微小的發現以提高對此病的病理學知識。自發性顱內低壓在香港並不常見，醫生必須在臨床上保持高度警惕，以便及早為病人診斷和進行適當的護理。此外，對於那些具復發性症狀或對治療無反應的病人，須考慮新的治療方法。

of the dura (spontaneous intracranial hypotension [SIH]), though a trivial traumatic event can be elicited retrospectively in around one third of such patients.[3,4] The most common sites of leakage identified were at the cervicothoracic junction and thoracic region of the spinal canal.[1,5] In the absence of a dural defect, a recent alternative hypothesis proposes increased CSF absorption from negative pressure gradient in the inferior vena cava.[6] In both instances, CSF hypovolaemia is the main feature and primary cause of the related clinical and imaging findings.

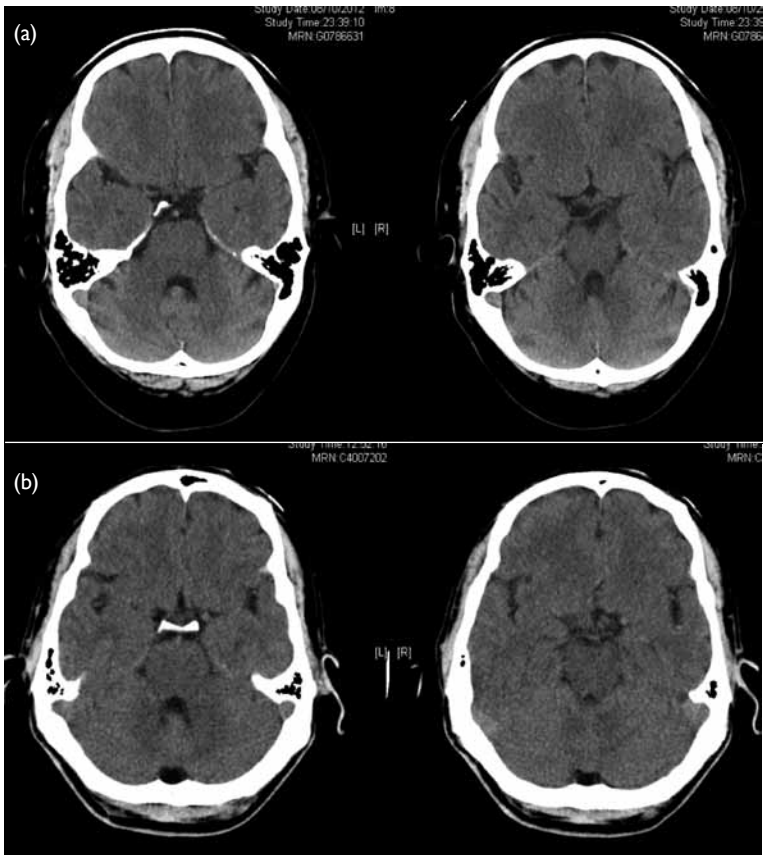

FIG 1. (a) Computed tomography of the brain (axial view) showing a normal brain with adequate space over the midbrain and basal cisterns. (b) Our patient's scan at similar axial level showing disproportionate tightness around the midbrain and basal cistern

Spontaneous intracranial hypotension is an increasingly diagnosed cause of headache, with an incidence of one in 50 000 individuals.[7] The female-to-male incidence ratio of SIH is 2:1, with a peak incidence occurring around the age of 40 years.[3,7]

The typical clinical feature of SIH is orthostatic headache, which, according to the International Headache Society, should occur or worsen within 15 minutes in an upright posture together with at least one feature of meningeal irritation (neck stiffness, tinnitus, hypacusia, photophobia, nausea) in addition to imaging features of SIH.[8] With chronicity, the postural component may become less prominent.[3] Other clinical manifestations include disappearance of or improvement in the headache within 30 minutes after lying supine, and cranial nerve palsies related to traction from caudal displacement of the brainstem.[9] Severe midbrain compression may result in nigral dopaminergic dysfunction and manifest as parkinsonism.[9] Coma occurs from delayed decompression of brainstem descent.[4]

Subdural haematoma is a late finding and occurs in around 10% of patients with SIH,[3] commonly seen in males and those older than 35 years.[10]

Computed tomography is the frontline imaging workup for headache. Typically, SIH is not considered unless patients present with non-traumatic SDH in the setting of a normal clotting profile. The proposed mechanism involves rupture of bridging veins in expanding subdural hygromas which, in turn, results from brain sagging due to CSF hypovolaemia.

On CT, SIH in the absence of SDH can be easily interpreted as being unremarkable. With high level of clinical suspicion, however, subtle imaging features may suggest the diagnosis. For instance, the initial CTB of our index patient showed paucity of CSF for his age (Fig 1). On follow-up CTB, the tightness of the basal cisterns appeared rather disproportionate to the small amount of SDH. Thus, it may be possible to detect CSF hypovolaemia on CT if these subtle findings are sought and the diagnosis is borne in mind.

The MRI findings of SIH are well-described in the literature. This preferred modality of imaging depicts characteristic features which may obviate the need for lumbar tap.[4] Additionally, the cause or site of dural defect can be investigated.

The MRI appearances are mainly attributed to CSF hypovolaemia, or represent secondary reactive changes following the Monro-Kellie doctrine. Briefly, decrease in CSF volume prompts an increase in dural blood flow and causes venous engorgement.

The latter, when prolonged, incites surrounding fibro-proliferation that in turn accounts for diffuse dural thickening and intense enhancement with gadolinium on MRI. Such explanation is confirmed by meningeal biopsy showing proliferation of fibroblasts without inflammation.[11,12]

The primary feature of brain sagging is a very specific MRI finding in SIH. It is a collaboration of features, including decreased dimension of the suprasellar cistern, bowing of optic chiasma, flattening of the pons against the clivus, effacement of the perimesencephalic cistern and hindbrain herniation (Fig 2a).[11,12] A quantitative measurement of brain sagging has been described[10] although the degree of descent may be underestimated since patients are scanned in the recumbent position.

Secondary and less-specific signs of SIH are more readily appreciated but represent a later stage of the disease. Findings of MRI in the brain classically show diffuse pachymeningeal thickening which has a sensitivity of up to 94% (Fig 2b).[9] The venous distention sign may also be seen and is best appreciated in the mid-portion of the dominant transverse sinus[12]; on sagittal sections, the sinus, which normally adopts a concave or straight inferior border, bulges with a convex contour. Venous engorgement at the dura mater across the sella turcica could produce reactive hyperaemia and possible increase in size of the pituitary gland. In the spine, MRI findings mirror those of the brain with diffuse dural enhancement, engorgement of venous plexus and extrathecal CSF collection.[3]

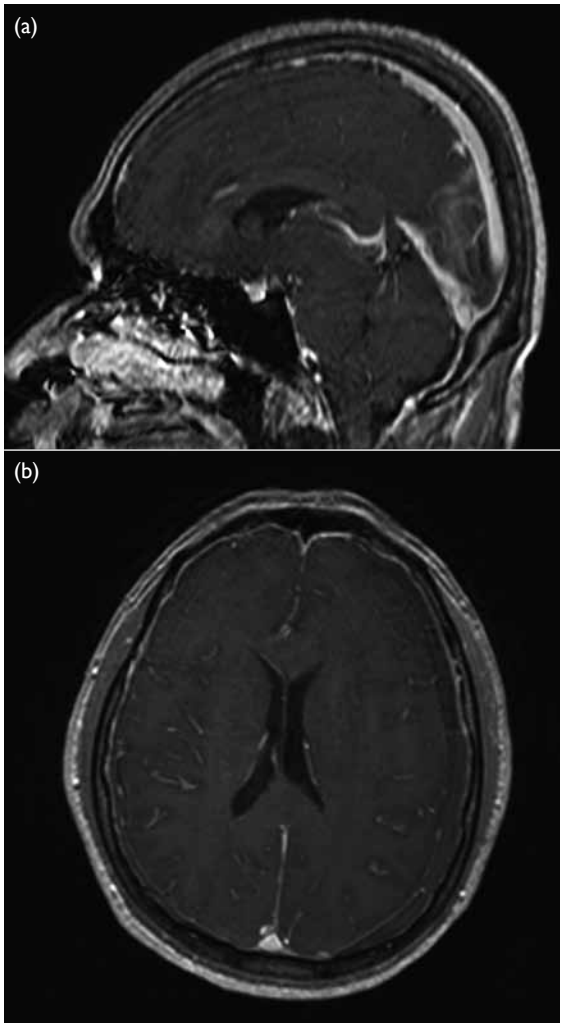

FIG 2. Magnetic resonance imaging of our patient showing typical features of spontaneous intracranial hypotension (SIH). (a) Brain sagging, best appreciated by the mammillary body, which normally lies on, but currently is, below the line between tentorial apex and tuberculum sella. (b) Classical diffuse dural enhancement seen in SIH

Magnetic resonance imaging cisternography or myelography can be easily added to routine MRI examination. Using a heavy T2-weighted sequence with fat signal suppression, spinal fluid outside the craniospinal axis may be detected with equal or improved accuracy than CT myelography.[5] Radiological improvement lags behind clinical recovery. Meningeal enhancement resolves considerably earlier than brain sagging.[12,13]

Computed tomography myelography and radionuclide cisternography are available locally but seldom performed. These are more invasive and time-consuming to perform, and entail intrathecal injection of contrast/radioisotope label, with fluoroscopic screening or serial imaging with gamma cameras to visualise extrathecal contrast/tracer activity.

Conservative management of SIH includes Trendelenburg positioning, aggressive hydration, caffeine intake and abdominal binder, and has been reported to be successful in most cases. In those patients with increased persistent headache or neurological deterioration, CTB should be immediately performed to rule out expanding SDH.

The timing of active surgical drainage is controversial. In a particular series, surgical drainage was advised in those with focal neurological deficits, decreased level of consciousness, or subdural collection of >1 cm.[4] Most surgically managed patients show transient improvement but have high likelihood of re-accumulation of subdural fluid.[13]

A more active strategy that is gaining international acceptance is epidural blood patch (EBP), which aims at sealing off the spinal leak[3] but appears to be useful in those without a definite dural defect.[6] The treatment involves placing 10 to 20 mL of autologous blood in the epidural space at the thoracolumbar level. As the patient is put in Trendelenburg position, the blood patch distributes along the epidural space and clots at the site of leakage.

The overall success rate for headache improvement is 30% to 70% after the first EBP and 30% to 50% for the remainder with repeated EBP.[10]

Epidural blood patch has a success rate of 85% in reverting patients who were comatose due to SIH.[13] Under fluoroscopic guidance, EBP may yield a high rate of pain relief and is preferred for those with altered anatomy or failure in the initial attempt.[14] A novel technique of multisite EBP via continuous infusion has been described.[15]

The newest treatment approach developed at the Stanford University recommends emergency subdural clot evacuation in the absence of improvement in Trendelenburg positioning, presence of dilated pupils, and large SDH with mass effect. Otherwise, EBP is the treatment of choice.[13]

In those with initial improvement with EBP, studies have shown concomitant spontaneous resolution of significant SDH.[13] Epidural blood patch may even be performed after surgical evacuation, and has been proven to further reduce the recurrence of headache and SDH.[13,16] As the brain has a tendency to sag downwards in SIH, pneumocephalus during clot evacuation may cause further downward herniation of the brain. Hence, surgical evacuation after EBP may not be advisable. Most of these patients, like our index case, have shown low-pressure subdural collection during craniotomy.

Conclusion

The diagnosis of SIH should be considered for any patient presenting with headache and neck pain. A high level of clinical suspicion could assist identification of subtle signs on initial CT which could facilitate early recognition and prompt treatment and, consequently, improve outcomes. However, MRI remains an important, highly sensitive and specific method for depicting imaging markers that allow confident clinical diagnosis. Further, MRI cisternography/myelography for CSF leak localisation can be added with ease. Currently, the medical practice in Hong Kong for SIH predominantly comprises conservative treatment and symptomatic clot evacuation. More novel interventions that have been successfully employed overseas may need to be reconsidered to improve current therapeutic strategies.

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