

SPONTANEOUS INTRACRANIAL HYPOTENSION RESULTING IN COMA: CASE REPORT

Faisal T. Sayer, M.D., M.Sc.

Department of Neurosurgery,
Lund University Hospital,
Lund, Sweden

Mikael Bodelsson, M.D., Ph.D.

Department of Anaesthesia and
Intensive Care,
Lund University Hospital,
Lund, Sweden

Elna-Marie Larsson, M.D., Ph.D.

Department of Radiology,
Lund University Hospital,
Lund, Sweden

Bertil Romner, M.D., Ph.D.

Department of Neurosurgery,
Lund University Hospital,
Lund, Sweden

OBJECTIVE AND IMPORTANCE: Spontaneous intracranial hypotension is a potentially severe condition characterized by a distinct clinical picture due to low cerebrospinal fluid pressure. Although coma has been previously reported as a presentation of this condition, this is the first report where misdiagnosis of this condition and unwarranted surgery lead to coma.

CLINICAL PRESENTATION: A 62 year-old man presented with a history of headache and cranial MRI showed bilateral chronic subdural haematomas. Following evacuation of the haematoma, the patient's condition deteriorated into a state of profound depression of consciousness. Repeated cranial CT showed intracranial air and intracranial pressure monitoring showed negative recording. Spinal MRI demonstrated epidural CSF leaks at the middle and lower thoracic levels.

INTERVENTION (OR TECHNIQUE): Epidural blood patch resulted in almost immediate improvement in the patient's condition and he was fully awake 24 hours later.

CONCLUSION: This case report expands the presently known clinical spectrum of this uncommon and generally benign illness.

KEY WORDS: brain sagging; CSF hypovolaemia.; spontaneous intracranial hypotension; subdural haematoma

Neurosurgery 58:E204-E205, 2006

DOI: 10.1227/01.NEU.0000219843.34356.EE

www.neurosurgery-online.com

Reprint requests:

Faisal T. Sayer, M.D., M.Sc.,
Department of Neurosurgery,
Lund University Hospital,
221 85 Lund, Sweden.
Email: Faisal.Sayer@neurokir.lu.se

Received, May 11, 2005.

Accepted, March 8, 2006.

Spontaneous intracranial hypotension (SIH) is an increasingly recognized cause of orthostatic headache, and it is a differential consideration in patients who present with nontraumatic spontaneous subdural hematoma. We document a case of SIH, where evacuating the associated chronic subdural hematoma was complicated by obtundation related to diencephalic compression. We discuss the appropriate diagnosis and management of SIH and demonstrate that the prognosis of the SIH seems good if appropriately diagnosed and treated in time before ensuing irreversible CNS damage.

CASE REPORT

A 62 year-old, previously healthy man, presented to his General Practitioner with three months history of headache, fatigue and progressive slowness in thinking and speech. The headache was described as dull, diffuse and it was accentuated by physical stress. There was no relation between the headache and posture.

He also complained of a mild short-term memory deficit of approximately the same duration. The patient also reported some degree of imbalance, but it was not sufficient to affect the gait and he did not need any support during walking. Further penetration of previous medical history revealed two months history of chronic cough, ascribed to allergy, approximately 6 months prior to presentation. There was no history of trauma or head injury. Radiological evaluation, within the same week of presentation, with MRI and CT scan of the brain showed extensive 10 mm thick bilateral fronto-parietal chronic subdural haematomas causing effacement of the sulci and compression of the lateral ventricles (*Fig. 1*). On MRI, a few mm descent of the tonsils below the foramen magnum was present (*Fig. 1a*), which was interpreted to be secondary to the mass effect of the haematomas. In addition, there was mild diffuse dural enhancement (*Fig. 1b*). Three days later, the patient was admitted to the department of neurosurgery at Lund University hospital on (Day 1). The patient

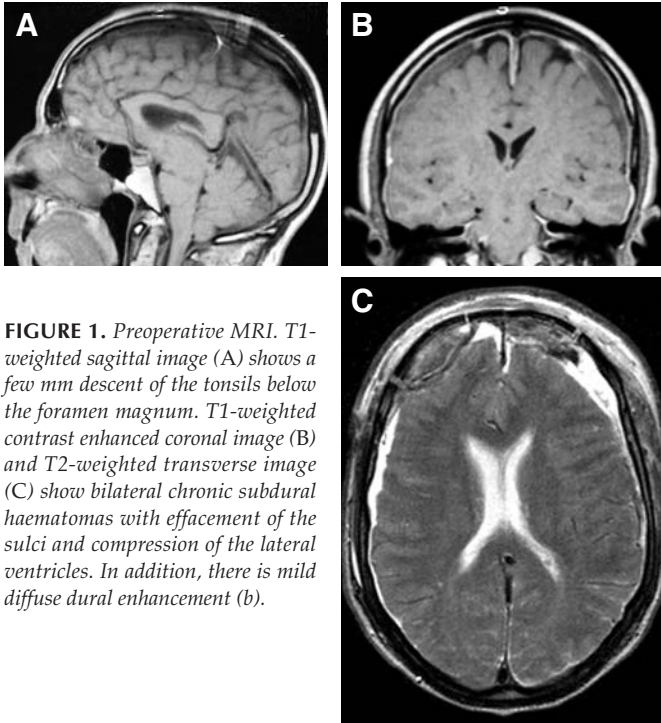


FIGURE 1. Preoperative MRI. T1-weighted sagittal image (A) shows a few mm descent of the tonsils below the foramen magnum. T1-weighted contrast enhanced coronal image (B) and T2-weighted transverse image (C) show bilateral chronic subdural haematomas with effacement of the sulci and compression of the lateral ventricles. In addition, there is mild diffuse dural enhancement (b).

displayed GCS 13 and answered questions correctly but with latency. Romberg's sign was positive with tendency to fall backwards. No other neurological deficits were found on admission. Standard physical examination, including blood pressure and pulse, as well as blood chemistry was normal. The following is a chronological outline of events during the patient stay at our department:

On Day 2, the patient was operated with burr hole irrigation (evacuation) of the chronic subdural haematoma on the left side. In the initial postoperative period, the patient recovered uneventfully. However, on Day 4, the patient was tired again (GCS 13) and a new CT scan showed intracranial air frontally ipsilateral to the surgical evacuation (Fig. 2a). The patient was kept under observation and his level of consciousness fluctuated between GCS 12 and 14. A new CT scan on Day 6 showed no increase of the intracranial air, however, the patient deteriorated further to a semi-comatose state (GCS 8). He was transferred to the neurosurgical ICU and an acute EEG the next day showed no evidence of epileptic activity.

On Day 8, the patient was re-operated with evacuation of the intracranial air through the same burr hole on the left side. Initially the patient's condition improved and he was more awake (GCS 14), but relapsed after 2 hours to GCS 8. A new acute CT scan showed an increased amount of intracranial air on the left side and the patient was re-operated the same night with evacuation of the intracranial air through the same burr hole. However, after this third operation, the patient remained in a GCS 8 state.

The following day, a new CT scan showed even more intracranial air in the bilateral frontal regions, and this intracranial

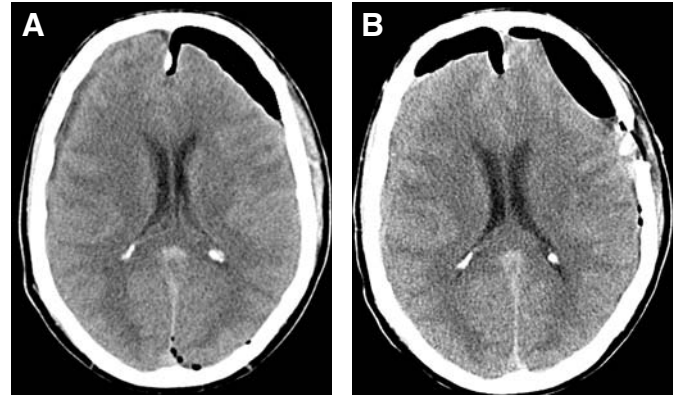


FIGURE 2. CT after the first operation (A) shows intracranial air frontally ipsilateral to the surgical evacuation. CT after the third operation (B) shows an increased amount of intracranial air, now in the bilateral frontal regions with mass effect on the frontal lobes.

air accumulation caused significant mass effect with about 2 cm dorsal displacement of the frontal lobes (Fig. 2b). The patient was operated acutely on the same night with a bi-frontal craniotomy, evacuation of the intracranial air and inspection of the floor of the anterior fossa for any possible source of air leak. No source of leak was identified. The patient's condition was unchanged during the following days (GCS 8).

An MRI examination of the brain on Day 12 showed a thin residual right-sided subdural haematoma and a left-sided subdural fluid-collection with effacement of the sulci of the convexities (Fig. 3). The previously noticed intracranial air had disappeared completely. In addition, this second MRI showed caudal displacement of the mesencephalon, pons and (Fig. 3a). Some degree of compression of the pons against the clivus was seen. The cerebellar tonsils were also herniated through the foramen magnum with progression compared with the preop-

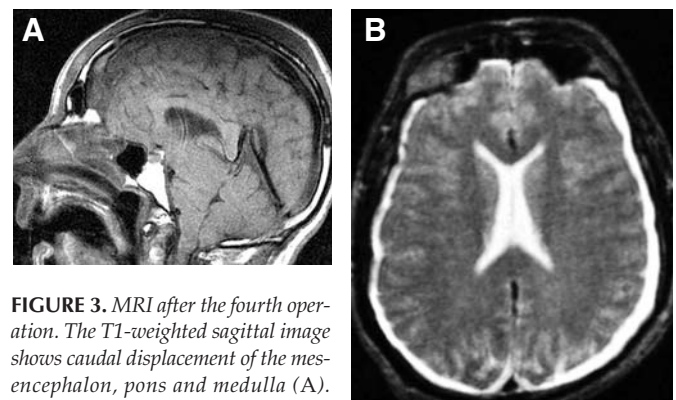


FIGURE 3. MRI after the fourth operation. The T1-weighted sagittal image shows caudal displacement of the mesencephalon, pons and medulla (A). Some degree of compression of the pons against the clivus is seen. The cerebellar tonsils are also herniated through the foramen magnum with progression compared with the preoperative MRI. The T2-weighted transverse image (B) shows a thin residual right-sided subdural haematoma and a left-sided subdural fluid-collection with effacement of the sulci of the convexities. The intracranial air has disappeared.

erative MRI. There were no signal changes in the brain stem that could explain the deterioration of the level of consciousness.

On Day 13, the patient's condition deteriorated further to GCS 7 and he was intubated and a catheter was inserted into the lateral horn of the right ventricle to monitor the intracranial pressure. When the ventricular catheter punctured the lateral ventricle, the CSF came out clear and under low pressure. The continuous registration of the intracranial pressure was always below zero (-50 to -90 mm H₂O). Because of the negative intracranial pressure and the MRI findings, spontaneous intracranial hypotension (SIH) was suspected.

On Day 15, whole spine MRI revealed an extradural fluid collection ventral to the spinal cord at C6-T1 and a similar collection dorsal to the spinal cord at the levels T1-T12 (Fig. 4). This fluid collection had the same signal intensity as CSF. No definitive source of CSF leak was identified. The clinical picture, negative ICP and radiological findings confirmed the diagnosis of SIH.

An epidural autologous blood patch (30 ml) was placed, under x-ray guidance after injection of 3 ml of contrast, in the epidural space at the levels L3-L4. Approximately 1 hour after placing the blood patch, the ICP started to increase and about two hours later, the ICP reached 140 mm H₂O and the patient was awake and obeying orders (GCS 13). The patient was kept sedated until the next day and extubation the next morning was uneventful. The patient quickly regained full consciousness. No residual neurological deficit was observed. On Day 21 the patient was sent to the referring hospital and was thereafter discharged home.

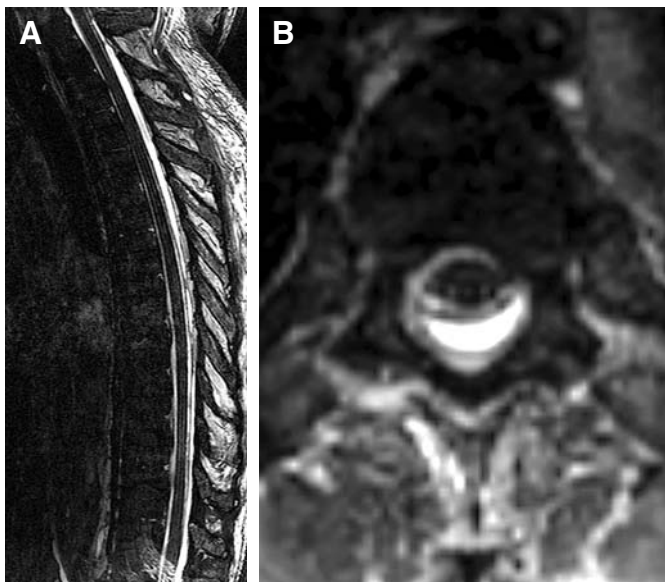


FIGURE 4. MRI of the thoracic spine after the fourth operation. 3D sagittal (A) and transverse (B) gradient echo images reveal an extradural fluid collection ventral to the spinal cord at C6-T1 and a similar collection dorsal to the spinal cord (arrows) at the levels T1-T12.

DISCUSSION

In 1938, Schaltenbrand described a syndrome of spontaneous positional headache with neck stiffness, nausea, vomiting, tinnitus, and vertigo in patients found to have low CSF pressure. Schaltenbrand called this condition spontaneous aliquorrhoea and postulated that it can be due to occult dural tear, overabsorption of CSF, or decreased CSF production (26). The SIH is now a recognised syndrome characterized by postural headaches and CSF hypovolemia (5). The mean age of the patients in was 43 years with some degree of female predominance (7).

The most common symptom of SIH is insidious onset of orthostatic headache, which often evolves into chronic daily headache. The headache associated with SIH is probably caused by

dilatation of the cerebral veins and meningeal vasculature, or it may be a consequence of the displacement of pain-sensitive structures secondary to low CSF pressure; or both factors (27).

Other symptoms may include visual field deficits, transient visual obscuration, facial numbness, sixth-nerve palsy, vertigo, tinnitus, nausea, vomiting, and rarely stupor, cervical myelopathy, and Parkinsonism (5, 19, 22, 23). Progressive personality and behaviour changes with memory loss may also occur (12). Although uncommon, coma has been reported as a presentation of SIH (7, 9, 18, 31).

The diagnosis of SIH requires a high degree of suspicion. A documented low CSF pressure (60 mm H₂O) in the absence of history of dural injury or tear (such as following lumbar puncture, back trauma, or operative procedures) is mandatory to establish such diagnosis (25). When SIH is suspected, MRI of the brain and spine are the investigations of choice. If MRI of the spine is normal, radioisotope cisternography and/or CT-myelography are performed to detect CSF leakage.

MRI Findings

Typical brain MRI findings suggestive of SIH include:

- 1) Subdural effusions (10% of cases) due to rupture of bridging veins caused by the decrease in CSF volume and downward displacement of the brain (10).
- 2) Diffuse thickening of the meninges (also known as pachymeningitis) and meningeal contrast enhancement (3).
- 3) Obliteration of basilar cisterns, descent of midline structures and flattening of the pons against the clivus (16).
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- 4) Mild tonsillar descent is one of the cardinal signs of SIH (18).

The dural enhancement is the most common and probably the earliest MRI manifestation of S.I.H., while subdural fluid collections and descent of the brain are indicators of a more severe hypotensive state (2).

Whole spine MRI may show varying degrees of shrunken dural sac, expanded extradural space with CSF accumulation and/or delayed homogeneous enhancement, and distended epidural venous plexus (4). In the majority of cases, no structural

lesion as a cause of CSF leak is identified and small tears in nerve root sleeves are considered responsible. Although MRI may not show the direct site of the leak, the location of the spinal fluid collection may correspond to the region of CSF leakage (i.e. cervical, thoracic or lumbar) (24).

Radioisotope Cisternography

Radioisotope cisternography with technetium-99m-human serum albumin is useful to document and localize spontaneous cerebrospinal fluid leakage along the neuraxis (13).

CT Myelography

On 3D CT myelography, the site of leakage may be depicted, which may assist targeted epidural blood patch under the guidance of fluoroscopy (10).

The choice of treatment will depend on whether a documented CSF leak is found or not. If no specific CSF leak is found, then a short course of steroid medications, orally administered caffeine and epidural or intrathecal saline infusion may be considered (1,20). If this has no effect or if the patient deteriorates, then epidural blood or fibrin glue patch should be employed as an attempt to seal the CSF leak (14, 17, 30). If these conservative measures fail and a dural defect have been demonstrated to be the underlying cause of the CSF leak, surgical repair of the defect should be considered. The follow-up of our patient, as well as the accumulated literature, indicate that a subduro-peritoneal shunt, used by Whiteley et al. (31), plays no role in the management of SIH. Although a patient death was reported previously (11), the prognosis of the SIH seems good if appropriately diagnosed and treated in time before ensuing irreversible CNS damage.

In our patient, the diagnosis of SIH was not made until he had been operated upon with evacuation of the subdural effusion and subsequently been reoperated three times with evacuation of intracranial air. This case should illustrate the value of establishing a list of differential diagnosis even in a seemingly routine and straightforward case like chronic subdural haematoma. Another lesson is that one should not be dogmatic about the diagnosis and this should be strongly discouraged since it can lead to disastrous consequences.

In retrospect, the initial MRI should have given some suspicion of intracranial hypotension. However, the subdural effusions with mass effect on the lateral ventricles were considered to be the main finding. The colour and consistency of the fluid evacuated at the time of initial surgery suggested that it was a hygroma and not a chronic subdural haematoma. Another observation was that when the dura was opened, the evacuated fluid was not under high pressure. These observations were neglected initially and this has contributed to the delay in establishing the diagnosis of SIH.

The low intracranial pressure, later proven by intracranial pressure monitoring, could explain this accumulation of intracranial air. Managing the patient in flat position for 24 hours after surgery did not reduce the intracranial air accumulation. Our patient had no clinical history of trauma, but his

chronic cough may have caused a dural tear with subsequent CSF leakage (21). The state of altered level of consciousness that the patient displayed was probably due to downward displacement of the brain and compression of the brain stem against the clivus. The fact that the patient showed dramatic and rapid clinical response to the lumbar epidural blood patch could indicate that the redistribution of the blood in the epidural space is faster than expected.

CONCLUSION

Early diagnosis of SIH is important in order to avoid unnecessary and potentially harmful evacuation of the associated subdural effusions. A straightforward case such as bilateral chronic subdural haematomas can turn into a nightmare if the history and radiological examination is not reviewed thoroughly before surgery.

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