

Spontaneous intracranial hypotension

Introduction

Spontaneous intracranial hypotension (SIH) is a rare headache syndrome associated with significant headache morbidity, cranial nerve palsies, subdural collections and haematoma, and even coma (O'Carroll and Brant-Zawadzki, 1999). Described by Schaltenbrand in 1983, the diagnosis can be made by the characteristic history of postural headache provocation with relief on recumbency. Over time such features may become less pronounced and the presentation evolves to one of chronic daily headache. Diagnosis is facilitated by char-

acteristic magnetic resonance imaging (MRI) changes (Pannulo et al, 1993). The authors present a case of SIH and a review of the clinical, radiological, and pathological features of this condition.

Discussion

Orthostatic headache is the most common clinical feature of low CSF pressure. It is a common iatrogenic complication of lumbar puncture (LP) but may occur in other scenarios as illustrated by this case.

SIH is defined by the absence of LP or other overt cause of CSF leakage. The aetiology of SIH remains obscure, but the

most common mechanism is believed to be as a result of the rupture of spinal dural diverticuli in the region of the cervicothoracic vertebrae. Other sites include the cribriform plate.

The duration of headache may vary greatly and can extend to several years. Associated features include nausea, vomiting, tinnitus, vertigo, cranial nerve palsies and local back pain.

Headache results from displacement of dura and vessels bearing nociceptive trigeminal sensory fibres. This is associated with secondary vascular changes as a result of the release of vasoactive compounds such as calcitonin-gene related peptide, and this underlies the diffuse meningeal enhancement.

The MRI findings of pachymeningeal enhancement with sparing of the leptomeninges are characteristic (Chung et al, 2004). Displacement of the brain may be evidenced by subdural collections and haematoma, and there may be incisural or cerebellar tonsillar herniation – the latter of which may be mistaken for a Chiari malformation (Good and Ghobrial, 1993). Radionuclide cisternography or contrast myelography may be helpful in refractory cases to identify the location of CSF leak (Molins et al, 1990). Early appearance of radionuclide in the bladder following CSF injection also provides clues to the presence of a dural leak.

Symptoms of SIH may be self-limiting. Conservative management includes bed rest, analgesia, antiemesis and rehydration – intravenously if necessary. Caffeine was demonstrated to be very effective in a double-blinded prospective study by Schzer

and Abel (1978). This study demonstrated benefit in 75% of subjects receiving 500 mg of caffeine intravenously. A second dose after 2 hours increases response to 85%. Its mechanism of action is unclear, but may involve adenosine antagonism-mediated vasoconstriction.

If symptoms are persistent a blood patch is indicated if the anatomical location of the leak can be identified. Its success rate may be as high as 97%. It is performed by the infusion of 10–20 ml autologous blood in the epidural space at the level of the leak under aseptic conditions. Rarely multiple diverticulae may be present and this may necessitate surgical intervention.

Conclusions

The syndrome of SIH can be diagnosed from its characteristic history and specific imaging changes. It may be a persistent debilitating condition but may respond to a range of therapeutic options from bed rest with fluids, to caffeine intravenously and rarely may require an epidural blood patch or surgical leak closure. **BJHM**

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Figure 1. Subdural effusions.

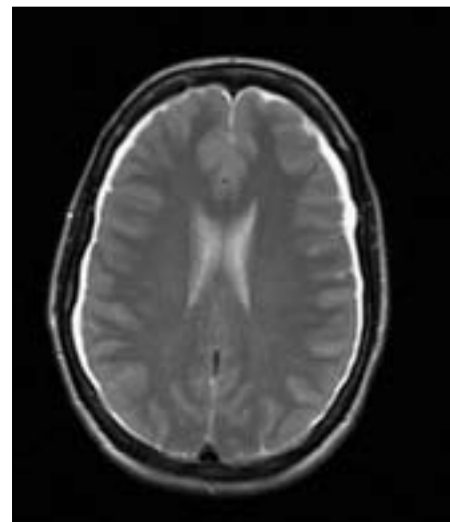
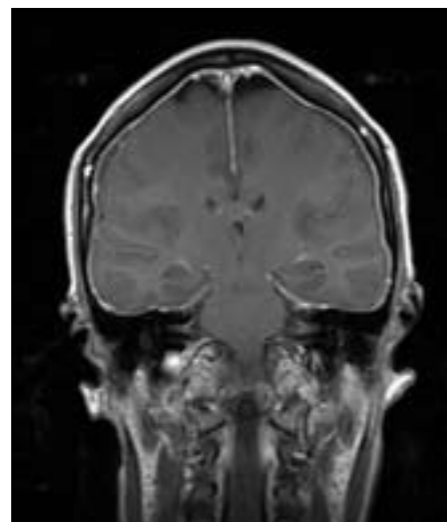


Figure 2. Pachymeningeal enhancement.



Case Report

A 47-year-old woman presented with a 4-week history of headaches. These were present from waking, and the preceding night's sleep had been disturbed by a sudden popping sensation associated with tinnitus. She described pounding frontal headaches on sitting or standing which were associated with nausea and vomiting, but relieved with bed rest. She had a past history of migraine without aura, which were qualitatively different, lacking a postural component associated with sensory sensitivity (photophobia and phonophobia). There was no history of trauma.

Detailed neurological examination was normal. There was no evidence of nasal or auditory meatal CSF leakage. Magnetic resonance imaging of brain demonstrated bilateral subdural effusions and there was diffuse enhancement of the pachymeninges following gadolinium (Figures 1 and 2). Magnetic resonance imaging (MRI) of cervical spine showed a small extradural collection of CSF density at the level of C6.

The patient was treated with bed rest and intravenous caffeine sodium benzoate (500 mg). Her headaches reduced in severity post caffeine such that she was able to sit and mobilize. Caffeine gave immediate but temporary benefit. Her headaches resolved completely over a further 4 weeks and a repeat MRI showed resolution of her collections and gadolinium enhancement.

At 6-month review she remains free of postural headache.

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