Case 3265

Spontaneous intracranial hypotension -- an important cause of postural headache
Published on 21.07.2005

DOI: 10.1594/EURORAD/CASE.3265
ISSN: 1563-4086
Section: Neuroradiology
Technique: CT
Technique: MR
Technique: MR
Technique: MR
Case Type: Clinical Cases
Authors: Kuriakose J, Kakkuzhi R, Azeez M, Widdowson D
Patient: 47 years, female

Clinical History:

A 47-year-old female patient presented with a postural headache and subdural collections.

Imaging Findings:

A 47-year-old woman with a history of migrainous headache for 20 years was referred to our hospital complaining of a severe headache of a month’s duration. She reported that the pain was different from her usual migraine attack. It was markedly postural, worse on sitting and standing and partially relieved by bed rest. There was no improvement of symptoms with the use of NSAIDS or anti-migraine medications. She had noticed an intermittent hearing loss, tinnitus and neck pain. There was no history of a recent trauma. An examination did not reveal any focal neurological deficits; there was no neck stiffness and the otological examination was normal. Routine blood tests including ESR and coagulation screen were normal. Computed tomography of the head (Fig. 1) revealed small bilateral subdural collections. Magnetic resonance imaging confirmed the presence of the subdural collections, showing a diffuse enhancement of the pachymeninges (Figs. 2 and 4) and an evidence of brain ‘sagging’ (Fig. 3). Based on these findings, a diagnosis of spontaneous intracranial hypotension (SIH) was made. The patient responded to conservative management including the use of IV caffeine administration, and made a full recovery. A repeat MRI examination of the brain and spine, after three months, was normal apart from a shallow residual left subdural collection with no evidence of brain sagging.

Discussion:

Spontaneous intracranial hypotension, first described by Schaltenbrand in 1938, is a benign condition characterized by intracranial hypotension which occurs as a result of reduced CSF volume, in the absence of lumbar/dural puncture, surgery or trauma. The pathogenesis is thought to be due to occult CSF leaks caused by dural tears, meningeal diverticula, and disc herniations. Fragile arachnoid cysts which are seen in association with occult connective tissue disorders have also been implicated. The commonest site of the CSF leak is in the cervico-thoracic junction followed by the thoracic spine, although the skull base or any part of the spine may be involved. As a result of the CSF hypovolemia, there is compensatory hypertrophy of the pachymeninges with dilatation of the blood vessels within, to maintain constant intracranial volume (Monro–Kellie hypothesis). The typical presenting symptom is an orthostatic headache similar to a post lumbar puncture headache, which as a result of the downward displacement of the brain, causing traction on the pain sensitive structures. The associated symptoms include neck
pain, photophobia, diplopia, tinnitus, facial numbness, dysguesia and changes in hearing, due to stretching of the lower cranial nerves. Other rare manifestations include Parkinsonism, dementia and coma from diencephalic or hind brain herniation due to severe brain sagging (1). CSF pressure is often less than 60 mm H2O and occasionally results in a dry tap, although normal pressures are also well recognized. The investigation of choice for the diagnosis of SIH is the magnetic resonance imaging of the brain. The characteristic imaging features which are seen are diffuse intense enhancement of the pachymeninges, the downward displacement of brain and subdural fluid collections. Meningeal enhancement was the commonest finding occurring in all 16 of the patients who underwent MR imaging, in a recently published series of case studies of 22 patients (2). The enhancement of the dura is thought to be due to dilated blood vessels. The leptomeninges are typically spared due to tight junctions in their microvasculature. Brain sagging is a specific finding in this condition, evidenced by the effacement of the suprasellar cistern, bowing of the optic chiasm over the pituitary fossa and flattening of the pons against the clivus—all three of which are seen in the case presented. The displacement of cerebellar tonsils, if present, can be mistaken for type1 Arnold Chiari malformation. Subdural fluid collections are seen in 10% and are typically bilateral, thin, and often without any mass effect. Other findings include hyperaemia of the pituitary gland mimicking a tumour. There is almost complete resolution of abnormalities seen on MR images obtained after treatment, paralleling clinical improvement (3) although meningeal enhancement may persist for more than three months after the disappearance of the symptoms. The most serious complication of SIH is subdural haemorrhage (SDH). Bilateral or unilateral SDH without obvious predisposing factors such as bleeding diathesis, trauma, alcoholism or anticoagulant medications, should suggest the possible diagnosis of SIH. Subdural haemorrhage is caused by the rupture of bridging veins as the brain sags, pulling away the dura. This complication was thought to be rare, with only a small number undergoing surgical SDH evacuation. However, SDH was present in four of the nine patients, in one case series, and all required surgical intervention (4). Hence, patients with diagnosed SIH should undergo a CT scan of the brain in the event of worsening symptoms or neurological deficits. Localization of the CSF leak is imperative only in patients who do not respond to conservative treatment. If surgical intervention is required for the evacuation of a subdural haematoma and the underlying spinal CSF leak is not corrected, the recurrence rate of SDH is high. The test of choice to localize leaks has been the CT myelography (CTM) technique, with spinal MRI and radionucleotide cisternography (RC) as guides for targeting the CTM to appropriate levels. The CTM procedure has a sensitivity of 67% compared to 50% and 55% of spinal MRI and RC respectively. A spinal MRI may demonstrate significant dilatation of the anterior internal vertebral venous plexus in up to 90% of the cases, as well as subdural hygromas, circumscribed retrospinal fluid collections between C1 and C2, dilated peri-neural sheaths and meningeal diverticula. It is capable of demonstrating both active and inactive leaks but has low sensitivity as it is dependant on patient's body type and level of the leak (5). An engorged superior orbital vein on Doppler-flow imaging is a highly specific and sensitive indicator of SIH. 75% of patients respond to conservative management with bed rest, hydration and oral or IV administration of caffeine. Lumbar epidural blood patches may be tried as treatment in case of the failure of conservative treatment. If the patient fails to respond to this therapy, then imaging is required to localize the leak for a more direct/closer blood patch. A procedure of continuous epidural saline infusions has been tried, to maintain the intracranial volume. Surgery is reserved for those patients in whom all the above measures fail. The ligation of the meningeal diverticula can achieve 100% resolution of the symptoms. In conclusion, the diagnosis of spontaneous intracranial hypotension should be considered in those patients with a positional headache and/or evidence of bilateral or unilateral SDH without obvious predisposing factors. Patients with SIH should be advised to seek urgent medical advice if their symptoms deteriorate and should undergo prompt cranial CT imaging to avoid complications.

**Differential Diagnosis List:** Spontaneous intracranial hypotension (SIH).

**Final Diagnosis:** Spontaneous intracranial hypotension (SIH).

**References:**
Figure 1

Description: An unenhanced CT showing shallow bilateral subdural fluid collections. Origin:
Description: An MR coronal FLAIR image showing bilateral subdural collections appearing hyperintense relative to the CSF. Origin:
Description: An MRI unenhanced sagittal T1-weighted image showing brain descent. Note the flattened pons, bowed chiasm and effacement of the suprasellar cistern. Origin:
Description: An MRI enhanced coronal T1-weighted image showing a diffuse enhancement of the meninges. Origin: