Spontaneous Intracranial Hypotension – Diagnosis and Management

Introduction

Spontaneous intracranial hypotension (SIH) produces a headache similar to a post lumbar puncture headache. Although the terms 'hypotension', 'low CSF pressure' and 'low CSF volume' are often used interchangeably, the syndrome can occur in the setting of 'normal' CSF pressures. Loss of CSF volume rather than pressure better explains the clinical features and imaging abnormalities.[1]

Classification of low volume headaches

The international classification of headache disorders recognises three subsets of low CSF volume headache.[2] These are post-dural (post-lumbar) puncture headache, CSF fistula headache and headache attributed to spontaneous low CSF volume/spontaneous intracranial hypotension. Criteria needed for these diagnoses are summarised in Table 1. The cardinal feature indicating low CSF volume headache is aggravation of symptoms within 15 minutes of sitting or standing. In post-dural puncture headache symptoms should improve within 15 minutes of lying down.

Aetiology and risk factors

SIH has been associated with abnormalities of the cervical spine and chiropractic manoeuvres. Although there may be a clear trigger such as a Valsalva manoeuvre or trauma, SIH may occur without a clear precipitant. The cause is thought to relate to a breach of dural diverticulae, or tearing of nerve root sheaths. Connective tissue disorders have been suggested as possible predisposing factors, as SIH has been reported in patients with Marfan's and Ehlers-Danlos syndromes.

Not all patients with spontaneous intracranial hypotension have low CSF pressures when measured at lumbar puncture. This implies that there may be significant individual variation in CSF pressures, and also that that the rate of CSF loss may be more important in producing the syndrome than the residual CSF pressure or volume. CSF leaks are not identified in every case of apparent SIH. In part this may relate to the limitation of investigations. However patients with a typical history, without clear evidence of a leak, and who have failed 'blind' blood patches, often prove very difficult to treat. It is possible in such cases that a CSF leak may have occurred, with a residual effect on CSF dynamics (eg lowered pressure setting in the choroid plexus, and sensitisation of meningeal afferents).

Based on similarities between post lumbar puncture headache (PLPH) and SIH, a number of inferences can be made. PLPH is less common at the extremes of age and this has been attributed to reduced epidural distensibility in the very old and very young.[3] A higher risk has been reported in young females with a low body mass index.[4] Patients with dementia appear to have a very low risk of PLPH[5] and this has been attributed to low pain sensitivity, rigid dural fibres, arteriosclerotic vessels, and large CSF spaces due to cerebral atrophy. PLPH occurs less frequently in those with higher CSF opening pressures.[6] These factors may also be of relevance in SIH.

Pathophysiology

Two main theories have been proposed to explain the cause of headache in patients with low CSF volume headache. They have been outlined in greater detail by Paldino et al.[7]

1) Traction on pain sensitive structures

Under normal conditions, CSF supports the brain reducing its weight from 1500g to only 48g within the cranium. This remaining weight is supported by suspension from several pain-sensitive structures. These include the meninges, cerebral and cerebellar veins (tributaries of the sagittal and transverse sinuses, respectively) as well as the fifth, ninth, and tenth cranial nerves and the superior three cervical nerves. Descent of the brain and traction on these structures, explains the orthostatic nature of the headache.

However tonsillar descent is not found in all patients with SIH. This may be because displacement of the brain is underestimated (because the patient lies supine during brain imaging). It may also be because there are additional pathophysiological mechanisms.

2) Dilation of pain sensitive intracranial vascular structures

The mean recumbent CSF pressure is approximately 150mm of water at all levels. In the erect posture, a pressure gradient occurs; highest in the lumbar sac, about 0 at the level of the cisterna magna, and around -85mm H_2O in the ventricles. Venous engorgement in both brain and spine occurs in SIH. According to the Monro–Kellie doctrine, the upright posture should be associated with further dilation of pain-sensitive intracranial venous structures. In support of this theory is the finding that coughing or Valsalva manoeuvres (that decrease the venous return to the heart and therefore increase intracranial venous volume) can reproduce headache in a patient with SIH even when supine.

Clinical Features

The onset of headache following SIH may be gradual or subacute but a thunderclap form is also well recognised in about 14% of cases.[8] Associated clinical features are neck stiffness, tinnitus, hyperacusis, photophobia, nausea, interscapular and radicular upper limb pain, vertigo, visual field defects, and cranial nerve palsies.

As SIH becomes chronic, the postural aspect of the headache may become much less apparent, and an index event may not be recalled. SIH should therefore be considered in the differential diagnosis of new onset persistent daily headache.[9]

Rare presentations of SIH include sudden deafness, orthostatic tinnitus, rapid onset encephalopathy and coma (attributed to diencephalic compression resulting from brain descent), Parkinsonism, and chronic behavioural features suggestive of frontotemporal dementia.[10-12] Radiculopathy due to cervical epidural venous engorgement has also been associated with SIH.[13]

It is also important to note that orthostatic headaches have been described without CSF leakage as the major clinical manifestation of postural tachycardia syndrome (a disorder characterised by chronic orthostatic symptoms and a dramatic increase in heart rate on standing, but that does not involve orthostatic hypotension).[14]

Investigations

Investigation of patients with suspected SIH may help corroborate the diagnosis and identify the site of CSF leakage.

Magnetic Resonance Imaging (MRI)

MRI brain with contrast is the initial investigation of choice in suspected SIH. Meningeal enhancement is the earliest and most frequent feature, occurring in more than 80% of subjects; tonsillar descent is seen in more than

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Table 1: Diagnostic criteria for headaches attributed to low cerebrospinal fluid volume (from the International Classification of Headache Disorders[2])

Post-dural (post-lumbar) puncture headache
Headache that worsens within 15 minutes after sitting or standing and improves within 15 minutes after lying
One of neck stiffness, tinnitus, hyperacusis, photophobia or nausea
Dural puncture has been performed
Headache develops within 5 days after dural puncture
Headache resolves spontaneously within 1 week or within 48 hours after effective treatment
CSF fistula headache
Headache that worsens within 15 minutes after sitting or standing and improves within 15 minutes after lying
One of neck stiffness, tinnitus, hyperacusis, photophobia, nausea
A known procedure or trauma has caused persistent CSF leakage with at least one of the following:low CSF pressure evidence on MRI, evidence of CSF leakage on conventional myelography, CT myelography or cisternography and CSF opening pressure <60 mm H2O in sitting position
Headache develops in close temporal relation to CSF leakage
Headache resolves within 7 days of sealing the CSF leak
Headache attributed to spontaneous (or idiopathic) low CSF pressure
Diffuse and/or dull headache that worsens within 15 minutes after sitting or standing, with at least one of the following
One of neck stiffness, tinnitus, hyperacusis, photophobia or nausea
A known procedure or trauma has caused persistent CSF leakage with at least one of the following: low CSF pressure evidence on MRI, evidence of CSF leakage on conventional myelography, CT myelography or cisternography and CSF opening pressure <60 mm H2O in sitting position
Headache develops in close temporal relation to CSF leakage
Headache resolves within 7 days of sealing the CSF leak

40% and subdural hygromas/haematomas in about 17%.[15] The subdural effusions result from transudation from engorged venous plexuses. Such changes range from very striking to quite subtle. Engorgement of venous sinuses may also cause enlargement of the pituitary gland.[16]

It is important to note that a normal MRI brain is compatible with the diagnosis of SIH,[17] and also that pachymeningeal enhancement can occur in the setting of significant and proven CSF leaks in patients who are headache free.[18] A normal initial brain MRI in symptomatic patients however is predictive of poor outcome.[19]

Cervical MRI scans may show meningeal enhancement and a dilated internal vertebral venous plexus in 85% and spinal hygromas in up to 70% of cases.[20] The utility of spinal MRI for detecting the site of CSF leakage relative to other modalities (see below) is not clear.

Doppler Flow Imaging

This is predicated on the basis that the superior ophthalmic vein is a tributary of the cavernous sinus and it might therefore reflect the engorgement of the intracranial venous sinuses that occurs in this condition. Increased diameter and maximum flow velocity of the superior ophthalmic veins has been demonstrated in patients with SIH using transorbital colourflow Doppler imaging. One study suggests this technique has very high specificity and sensitivity, though clearly does not assist in identifying the site of leakage.[21]

Radionucleotide Cisternography

Radionucleotide cisternography frequently demonstrates 'surrogate' markers' of a low volume CSF state. These include limited ascent of the tracer to the cerebral convexity in 91% of cases, early appearance of the radioisotope in the bladder in 65%, and early soft tissue uptake of radioisotope in 43%.[22] Actual leakage of CSF has been identified in 52% of cases,[22] most commonly at the cervico-thoracic junction or in the thoracic spine. Intermittent leaks may go undetected and the technique may be insufficiently sensitive to identify small leaks.

CT Myelography

CT myelography has been found to demonstrate the level of a CSF leak in 67% of patients overall, compared with only 50 and 55% for spinal MR imaging and radionucleotide cisternography.[23] In no case did radionucleotide cisternography reveal the leak when CT myelography did not. Unfortunately CT myelography can be very time consuming, as it requires CT slices be obtained through the skull base and the entire spinal axis. Spinal imaging and radionucleotide cisternography may perhaps be helpful as guides for focusing on particular areas with CT myelography.

Lumbar Puncture

Lumbar puncture should be considered only if the features are equivocal and should be avoided before MRI with contrast as this can interfere with interpretation of the results. CSF opening pressure is typically low (usually 0–5cm CSF).[5] It may however be normal in up to 17% of cases.[24] CSF constituents are usually normal although high protein concentration and lymphocytic pleocytosis may be seen.[25]

Treatment

Medical

Conservative measures like bed rest are the first line treatment for low intracranial pressure headache. If not effective, intravenous caffeine at a dose of 500mg in 500ml saline over two hours (repeated once or twice) is often used although the evidence base is limited.[9] Cardiac monitoring is necessary as caffeine can induce arrthymias. There is also some evidence to suggest that theophyllines may be efficacious. It has been proposed that methylxanthines produce arterial constriction through the blockade of adenosine receptors.[26] Consequently, intracranial blood flow and, presumably, venous engorgement are decreased. Abdominal binding with a surgical corset may help, while glucocorticoids or mineralocorticoids have been used in some studies but are of questionable effect.

Interventional

i) Autologous epidural blood patch

The technique was initially based on the observation that PLPH was less severe after a 'bloody tap' compared with a 'clear tap'. The mode of action is not clear but may be due to a tamponade effect. It is performed by slowly injecting autologous blood into the same interspace or the interspace below the site of leak.

In contrast to PLPH the site of CSF leakage may not be certain in SIH. It may not be critical to identify the site of leakage. There is some evidence that lumbar epidural blood patching may be effective over nine spinal segments when the patient's head is lowered to 30°. A recent report suggests that early 'blind' epidural blood patching within one week of onset is effective; demonstrating complete cure in 77% of 30 patients (with or without typical MRI changes) after one (57%) or two (20%) blood patches. These patients did not have lumbar punctures, nor was the site of CSF leakage identified.[27]

ii) Other treatment modalities

Epidural saline injection has been reported to give immediate relief for headache. This is thought to be by reduction in the distensibility of the epidural space. This manoeuvre could also be life saving in obtunded patients with SIH.[28]

A small group of patients with a typical history but no clear evidence of a leak, may fail 'blind' blood patches. These patients often prove very difficult to treat. It is possible in such cases that a CSF leak may have occurred, with a residual effect on CSF dynamics (eg lowered pressure setting in the choroid plexus, and sensitisation of meningeal afferents).

Conclusion

Loss of CSF volume best explains the syndrome often designated 'low-pressure headache'. CSF pressures may not always be low.

Patients with chronic symptoms of SIH may not volunteer or recall a

definite ictus and over time the postural aspect of the headache may become less clear. SIH may thus present as new onset persistent daily headache rather than as an orthostatic headache. MRI brain with contrast is the first line investigation of SIH. Patients with a normal contrast enhanced MRI brain appear to have a worse prognosis. Radionucleotide cisternography generally shows abnormalities in SIH although it is relatively poor at locating the site of a CSF leak. CT myelography of the spine is arguably the most sensitive test to identify the site of a leak. However it is time consuming if there are no clues as to where to focus the study.

It is usual practice to try and focus a blood patch on the site of the CSF leak (the most common site is cervico-thoracic). However this may incur a delay to treatment while imaging investigations are arranged. Recent studies suggest that 'blind' blood patching at an early stage may be very effective.

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