Spontaneous intracranial hypotension and postural headache

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Spontaneous intracranial hypotension (SIH) is caused by a spontaneous leak of spinal cerebrospinal fluid. Orthostatic headache is the hallmark of this condition, but due to under-recognition by clinicians and varied presentations, delays in diagnosis are common. A high degree of suspicion is required to make the diagnosis, and once confirmed by neuroimaging, it can often be managed successfully with an epidural blood patch. Novel treatments for this condition include an epidural injection of fibrin glue, with surgical repair reserved for unresponsive patients. This case report illustrates the diagnostic challenge, investigation and management of this condition. It also demonstrates the importance of relevant clinical information when interpreting imaging, and finally, aims to raise awareness of this important clinical entity.

Case presentation

A 37 year old female social worker presented to the emergency department with a three month history of intermittent headache, recently increasing in severity and duration in the last six to seven days. The headache started in the cervical spine, and curved over the head and into the face bilaterally, with a sensation of ache and pressure. Associated symptoms included nausea, vomiting, unquantifiable weight loss and intermittent auditory symptoms. Assessment by an ear, nose and throat specialist including audiometry did not reach a diagnosis. Of note, the headache was worsened when standing upright, and relieved when supine. She gave no history of prior lumbar puncture, fever, visual disturbance, numbness, paraesthesiae, weakness, urinary symptoms, vertigo or tinnitus. She had presented to her general practitioner several times and underwent a magnetic resonance imaging (MRI) brain which was reported as unremarkable. Two days prior to this presentation she attended a regional hospital Emergency Department (ED), and the next day attended a tertiary hospital ED, where she was discharged for neurology follow up. As she was incapacitated by the headaches and was struggling to care for her children while her husband worked away from home, she presented to the private ED a day later.

She had a past medical history of anxiety and depression, with symptoms well controlled on sertraline 25mg daily, prior tension headache and a past surgical history of two caesarean sections. She was taking no other regular medications and there was no relevant social or family history.

On examination, she was a young woman in no apparent distress with normal vital signs. Peripheral and cranial nerve examination, and fundoscopy were unremarkable. There were no cerebellar signs. Cervical spine examination demonstrated tenderness on palpation on the mid spine of C1 and C2, with pain on flexion and right and left lateral rotation. The remainder of the examination was unremarkable.

Clinical findings were consistent with spontaneous intracranial hypotension (SIH). She was admitted to the ward and managed on simple analgesia (paracetamol, Panadeine® and Mersyndol Forte®), temazepam and ondansetron. In light of the diagnosis, MRI brain with gadolinium and magnetic resonance venography was repeated on Day One. This demonstrated features consistent with intracranial hypotension: inferior descent of the cerebellar tonsils through the foramen magnum (Chiari 1 malformation), reduction in the suprasellar cistern, bilateral shallow subdural hygromas over both convexities and mild diffuse dural enhancement within the supratentorial location. A magnetic resonance venogram demonstrated patent dural venous sinuses. MRI cervical spine demonstrated a small volume of fluid between the posterior elements of C1 and C2 which was extradural in location. Cervical spine degeneration was described without cord compression, and there was some mild foraminal narrowing bilaterally at C5/C6.

Admitted under the general physician, she was reviewed by the neurologist on Day Three, who recommended a MRI of the thoracic and lumbar spine. This demonstrated degenerative changes of discs but failed to identify the location of the leak. As such, a lumbar epidural blood patch was performed by the anaesthetist on Day Five. Follow up on Day Six was normal, and she was discharged with advice to contact her neurologist for any further issues. When followed up four months later, she was asymptomatic and satisfied with the outcome. She was also able to return to full activities.

Discussion

Intracranial hypotension is defined as an abnormally low intracranial pressure (normal 10-15mmHg), and is characterised by headaches exacerbated by standing and relieved when supine. [1,2] Spontaneous intracranial hypotension is caused by spontaneous leakage of cerebrospinal fluid (CSF) through the dura mater. Whilst it is an important cause of new headaches in young and middle-aged individuals, it is not well recognised, leading to misdiagnosis, significant delays or never being diagnosed. [3] Initially described by Georg Schaltenbrand in 1938, this eponym has fallen out of favour due to his role in the Third Reich. [1,4,5] The prevalence has been estimated at 1 per 50,000 headache presentations in an emergency department study, with a female preponderance (male to female ratio of approximately 1:2). [6] Symptoms typically occur in the 40s and 50s with a peak incidence around 40. [3]

The exact aetiology often eludes discovery, but two contributing factors that are frequently suspected include weakness of the meningeal sac in certain regions and trivial traumatic injury (in about one third of patients). [3,7] Other predisposing factors include individuals with underlying connective tissue disorders, such as Marfan syndrome, Ehlers-Danlos syndrome type II and autosomal dominant polycystic kidney disease, as these increase the likelihood of CSF leak from

spontaneous dural tears or the formation of meningeal diverticula. [3,8] Other causes of localised defects of the connective tissue, such as vertebral spurs, and dural rents and tears have been observed during surgery. [8] Symptomatic causes such as spontaneous CSF rhinorrhoea or otorrhoea, uraemia or diabetic coma are less common and the obvious clinical presentation facilitates the diagnosis. [1] Uncommon causes involving the piercing of the dura include osseous spinal pathology caused by congenital defects or acquired degenerative disc disease.[8]

Leak of CSF at a spinal level causes CSF hypovolaemia, resulting in a reduction in brain buoyancy. This causes sagging of the brain in the cranial vault and stretching of pain-sensitive structures, such as the venous sinuses around the brain, the blood vessels of the meninges and the dura mater, accounting for the characteristic postural symptoms. [8,9] Vascular congestion may also contribute to the headache. [8] Physiological compensation is explained by the Munroe-Kellie hypothesis, which states that the sum of volumes of brain, CSF and intracranial blood is constant. Thus a decrease in one should cause an increase in one or both of the remaining two. [10] In response to CSF hypovolaemia, there is an increase in the vascular component (by dilatation of compliant intracranial blood vessels) as the volume of brain parenchyma remains constant. [8] In addition, hygromas may form to restore depleted intracranial volume. Therefore CSF hypovolaemia has been proposed as the underlying mechanism for the headache syndrome, as opposed to CSF hypotension per se. [11]

The diagnosis of SIH is challenging due to the variability in headache symptoms and the lack of awareness by clinicians. This diagnosis should be considered in patients presenting with a postural headache or a daily persistent headache without an alternative cause. The hallmark of this condition is a postural headache that occurs or worsens within 15 minutes of assuming an upright position, and improves rapidly, usually within 15 to 30 minutes, of assuming a recumbent position. In some patients, however, the time period before the onset of the headache may be as long as several hours. Patients in the chronic phase may experience resolution of the headache but persistence of other symptoms. In rare cases, postural headache may not be experienced, even at the outset. In some, the headache becomes persistent rather than postural. [3,8,12,13]

The onset of headache may be gradual or sudden, and rarely starts as a thunderclap headache, which is seen in subarachnoid haemorrhage. It is often described as generalised and throbbing in character, but may be reported as dull or focal involving the frontal or occipital region. The severity of the headache varies from mild to incapacitating. Other exacerbating factors include head movement, straining, coughing, sneezing, jugular venous compression and high altitude. Other clinical manifestations include back pain, radicular symptoms, postural neck discomfort, nausea, vomiting, dizziness, vertigo, gait unsteadiness, ataxia, blurred vision, photophobia, transient visual obscurations and diplopia attributable to cranial nerve VI palsy. Hearing disturbance due to traction on the cochlear nerve or abnormal intra-labyrinthine pressure may cause tinnitus or hypoascusis (reduced sensitivity to sounds). [3,8,12,13]

Infrequently, more severe neurological manifestations occur, including Parkinsonism, quadriparesis and cerebellar haemorrhage. Pituitary congestion and enlargement may cause hyperprolactinaemia and galactorrhoea. Patients may also present with subtle cognitive deficits or coma. Notably, most of the neurological deficits are reversible following treatment. [3,8,12,13]

Neuroimaging, particularly MRI, is of great utility in establishing a diagnosis and identifying the site for later intervention. Major MRI findings with gadolinium enhancement include subdural fluid collections, enhancement of the pachymeninges with leptomeningeal sparing, engorgement of the venous structures, pituitary enlargement and sagging of the brain with cerebellar tonsillar herniation. Other findings include a decrease in the size of cisterns and ventricles, pituitary hyperaemia, swelling of the upper brain stem, effacement of the prepontine cisterns, descent of the cerebellar tonsils resembling Chiari 1 malformation, subdural haematomas (in up to 20%) and subdural hygromas. [8,13,14]

Computed tomographic (CT) myelography and Gadolinium-enhanced MRI may be used to identify the site of the leak. MRI scans may be normal in up to 20% of patients, and radionucleotide cisternography may be used to detect a leak instead. A brain CT is usually normal, but may demonstrate slit-like ventricles or tightness of the basal cisterns, and subdural hygromas. Prior to the recognition of characteristic MRI findings, lumbar puncture was the study of choice. Findings included low CSF opening pressure, but this could appear normal despite CSF hypovolaemia on neuroimaging. This may have been due to prolonged recumbency or an intermittent leak. Rarely, an inability to obtain CSF may be experienced. Other CSF studies are usually normal, but mildly raised protein and a reactive pleocytosis may be demonstrated. [8,13,14]

In one case series, 42% of patients managed conservatively (including bed rest, oral or intravenous hydration, analgesia and 200 to 300mg of caffeine orally twice to three times a day) recovered. [8,15] Adjuncts include abdominal binders (which may help some) and glucocorticoids (some benefit reported but it is yet to be proven). The duration of conservative treatment before the decision for other intervention is made on a case by case basis. [3,8]

Epidural blood patch (EBP) is currently the mainstay of treatment, and involves 10 to 20mL of autologous blood injected into the epidural space. [8] The success of the first patch varies (36-57%), with targeted epidural blood patch being more effective. [7,16] The mechanism of the EBP is believed to be the spread of blindly patched blood through the lumbar route. This may then spread to the cervico-thoracic level when the patient is in the Trendelenburg position, plugging the dural rent. The fibrin reaction at the site of the leak forms a scar in two to three weeks. A tamponade effect is provided and causes some degree of restoration in the CSF volume, resulting in symptomatic relief. Small retrospective studies and case series demonstrate that more than one EBP is required in at least 50% of cases. If the second patch fails, neuroimaging is used to visualise the leak and a targeted blood patch is applied. This has been shown to be more effective than blind EBPs. [8,16] A novel technique using a double EBP at two different levels (lumbar and thoracic) in the same procedure has been described for cases of SIH without a clear, demonstrable CSF leak. [17] Further studies are required to gauge the value of this technique.

Surgical repair is the last resort, and is reserved for patients who fail to respond to conservative management and blood patches. There is close to 100% success rate for meningeal diverticulae treated by simple ligation and repair of meningeal tears. [13] Novel treatment modalities include an epidural injection of fibrin glue for patients unresponsive to an epidural blood patch. Epidural saline infusion has also been attempted, producing immediate relief albeit with reduced efficacy. [13] Finally, patients with a long history of the disease may develop chronic headache syndromes. This pain is unrelated to posture, but is highly refractory to analgesia. [18]

Conclusion

Spontaneous intracranial hypotension is an important cause of new onset headache in young and middle-aged individuals. A detailed history and examination can alert the clinician to include this condition in the differential diagnosis. Subsequent investigation with neuroimaging provides a sensitive diagnostic test, and may locate the site of the leak. Treatment is often conservative or with an epidural blood patch, leading to complete resolution of symptoms in the majority of patients. SIH is frequently overlooked as a cause of headache. An increased awareness of this condition would provide earlier diagnosis and significant improvement in quality of life for these patients.

Consent declaration

Informed consent was obtained from the patient/next-of-kin for publication of this case report and accompanying figures.

Conflict of interest

None declared.

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