Spontaneous Intracranial Hypotension


Abstract

Spontaneous intracranial hypotension (SIH) is an increasingly recognised syndrome. Postural headache with typical findings on magnetic resonance imaging (MRI) are the key to diagnosis. Orthostatic headache, low cerebrospinal fluid opening pressure, and diffuse meningeal enhancement on post-contrast T1-weighted MRI brain studies are the major features of this increasingly recognised syndrome. Headache due to SIH is similar to headache occurring after lumbar puncture. Delay in diagnosing this condition may subject patients to unnecessary procedures and prolong morbidity. We describe a patient with SIH and outline the important clinical and radiographic features of this syndrome. Patients with postural headaches should have brain MRI before lumbar puncture. When correctly diagnosed, SIH management, in most cases, is easy and highly effective.

Introduction

Spontaneous or idiopathic intracranial hypotension is rare. Careful history taking and a high level of suspicion are essential to diagnosing this syndrome. Imaging studies, especially MRI, are helpful in confirming the diagnosis and ruling out any other conditions. This condition is usually treatable with simple measures, though failure to recognise it may subject the patient to unnecessary procedures, workup, and treatments. To familiarize physicians with SIH, we present the following case.

Case Report

A 24-year-old right-handed, previously healthy man presented to the Emergency Department after having daily headaches for 4 weeks. He described the headache as a dull ache of moderate intensity that persisted for several days in occipital regions. It was associated with neck pain and had become unbearable since 2 – 3 days. Pain began abruptly and peaked within 30 minutes of getting up in the morning. The onset of severe headache occurred initially while the patient was working as an office assistant. He described no aura, visual change, or neurologic symptoms. He described the headache as being relieved by assuming a recumbent position for a few minutes, only to be exacerbated by 30 minutes after assuming an upright position and doing his activities. There was no past history of headaches or precursors of migraine. There was no history of convulsion, blurring of vision, diplopia, photophobia, decreased hearing, skin rash, tick bites, head trauma, or fever or chills, vertigo, dizziness, unsteady gait and family history was negative for headaches. Patient took treatment from a private practitioner for 3 – 4 weeks without much relief. His past medical history was unremarkable.

On physical examination his vital signs were normal. Blood pressure was normal without postural hypotension, and he was afebrile. He was conscious and oriented. Ophthalmologic examination showed normal eye movement, equal, round reactive pupils and fundoscopic examination was also normal. His vision was intact. Ear
Discussion

Spontaneous intracranial hypotension (SIH) presenting with headaches is often misdiagnosed as migraines, tension headaches, or viral meningitis. The estimated annual incidence is 5 per 100,000. Women are twice as likely to have this condition compared with men with the onset usually occurring at age 40 to 50. SIH is caused by spontaneous spinal cerebrospinal fluid (CSF) leak. The cause of CSF leaks is often not known. Many patients with a CSF leak have been found to have connective tissue disorders including Marfa and Ehlers-Dandles syndromes. About one third of patients can recall some trauma, even if minor. It is postulated that minor trauma could cause rupture of spinal epidural cysts or per neural cysts.

The classic headache associated with SIH is an orthostatic headache that worsens or starts 15 minutes after sitting or standing. The headache in spontaneous intracranial hypotension may be gradual or acute in onset, and may localize to the frontal or occipital regions.

Usually the headache improves by lying down. Patients may describe the headache as either diffuse or localised often to the occipital region and it is almost always bilateral. Normally CSF supports the brain such that its weight of 1500 gm amounts to only 48 gm within the cranium. Depletion of CSF volume with downward displacement of the brain causes traction on the pain sensitive structures. The Monroe-Kelly hypothesis proposes dilatation of intracranial pain sensitive vascular structures as a cause of headache, which could be worsened in upright position and by Valhalla manoeuvre.

The most common associated symptoms are posterior neck pain or stiffness, nausea and vomiting. Hearing complaints are also very common and are believed to be caused by pressure changes that are transmitted to the cochlea. The sagging brain may also stretch the eighth nerve and other cranial nerves, which is felt to be the mechanism for visual changes such as blurred vision, diplopia, and photophobia. Other common symptoms are dizziness, vertigo, unsteadiness, and hiccups. The atypical features include Parkinsonism, front-temporal dementia, hypopituitarism, seizures, coma and death. The cause of this hypotension can be due to either CSF leak or can be spontaneous without any apparent cause.

Physical examination is usually normal, however patients can have abducens palsies or visual field defects. The lumbar puncture opening pressure usually is low ranging from 0-70 mmH2O.

The International classification of Headache disorders requires the following to diagnose a headache due to spontaneous spinal CSF leak: the orthostatic headache as described above along with at least one additional symptom (neck stiffness, tinnitus, hypoacusia, photophobia, or nausea) and confirmatory evidence of low CSF pressure such as on magnetic resonance imaging (MRI) with gadolinium, conventional myelography, computed tomography (CT) myelography, cisternography, or CSF opening pressure.

The most helpful study is a MRI of the brain with the...
following characteristics: subdural fluid collections, enhancement of the pachymeninges, engorgement of the venous structures, pituitary enlargement (hyperemia), and sagging of the brain. The meningeal enhancement has been described as continuous over the convexities, along the inter-hemispheric fissure and tentorium but not involving the depths of the sulci. Venous hypervolaemia is felt to be the cause for enhancement of the pachymeninges. Of note, it has been estimated that up to 20 percent of patients can have a normal MRI. If this is the case, CT myelography is recommended as it is the best at localising the site of the leak.

Although there have been no randomised clinical studies to evaluate treatment options, the initial treatment is either conservative treatment or an epidural blood patch which consists of autologous blood being injected into the epidural space. It is believed to work by sealing the leak and is effective in relieving symptoms in a third of patients. If this fails, it can be repeated. Other treatments such as a directed epidural blood patch or percutaneous placement of a fibrin seal require knowledge of the exact site of the CSF leak. Patients who fail these treatments should be referred for surgical repair. Despite treatment, some patients continue to have persistent symptoms.

References