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Case Report

Spontaneous intracranial hypotension: a case report in the context of differential diagnosis of chronic headaches

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Abstract

Spontaneous intracranial hypotension (SIH) is a rare and debilitating condition caused by decreased intracranial pressure, which occurs more frequently in females. SIH can have several causes, which is primarily responsible for the appearance of postural headache. Orthostatic headache is diagnosed by CSF pressure < 6 mmHg associated with specific imaging findings. Other specific symptoms such as dizziness, reduced muscle strength, blurred vision and syncope and other more systemic symptoms such as fatigue, mental confusion and difficulty concentrating are commonly observed. Etiological investigation through imaging studies such as magnetic resonance imaging and dynamic tomography of myelography is necessary for diagnosis. Due to the debilitating condition, several therapeutic approaches have been developed, ranging from more conservative approaches, with observation and use of analgesics, to more invasive interventions such as surgical ligation, transvenous embolization and blood tamponade.

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Introduction

Spontaneous intracranial hypotension (SIH) is a debilitating condition resulting from non-iatrogenic leaks of cerebrospinal fluid (CSF) in the spinal column, leading to decreased intracranial pressure.[1] It is a rare clinical condition, with an incidence of 1 in 50,000 subjects and more common in females.[1,2] The causes of SIH include ventral dural leak, spinal nerve root diverticulum with the leak, or direct venous fistula of CSF.[2]

The main features of SIH include a postural headache due to orthostatic hypotension caused by dilation of cerebral veins and meningeal vasculature, along with low CSF pressure.[3] It can also be associated with debilitating symptoms such as dizziness, muscle weakness, blurred vision, and syncope.[3]

The treatment of SIH involves targeted approaches to repair the CSF leak, depending on its location and extent.[4,5] Conservative measures such as horizontal bed rest, adequate hydration, and analgesics for pain relief may be considered.[4,5] In severe or refractory cases, surgical treatment may be necessary, such as surgical ligation, nerve root skeletonization, and blood patching of the CSF leak.[4,5]

Therefore, the present study aims to report the case of a patient with chronic headaches due to SIH caused by a posterior dural leak, treated with the correction of the CSF fistula using biological glue and an epidural blood patch.

Case report

A 17-year-old female patient presents with a 3-year history of persistent chronic headache. The headache is holocranial in presentation, characterized as constrictive, disabling, and resistant to treatment with topiramate 250 mg/day, Venlafaxine 75 mg/day, and chlorpromazine 4%, 12 drops/day. The condition worsens significantly when in an upright position and during physical exercise but provides brief relief when lying down. There were no reports of photophobia, phonophobia, aura, associated nausea, or vomiting. The general condition, neurological examination, and fundoscopy are unremarkable upon physical examination. There was no history of associated prior surgeries or lumbar puncture. The patient was then diagnosed with cranial pain syndrome involving the meninges, blood vessels, and pericranium. A cranial magnetic resonance imaging (MRI) was performed to further investigate the etiology due to the persistent headache with alarm signs. The cranial MRI showed no abnormalities. The diagnostic hypothesis of CSF hypotension was raised, and a neuroaxis MRI was requested to investigate further. The neuroaxis MRI revealed a possible CSF leak at the lumbar level, L4-L5. A myelography MRI was also requested, confirming a discontinuity of the dural sac on the right side, and intrathecal myelography sequences revealed a CSF leak in the topography of the L4-L5 CSF fistula. Therefore, the diagnostic hypothesis of the case became spontaneous intracranial hypotension secondary to CSF fistula due to "encysted" fluid within the posterior spinal canal, causing mild compression on the posterior aspect of the dural sac. As the chosen approach, laminectomy and correction of the CSF fistula were performed using biological glue and epidural blood patching. After the therapy, the patient showed significant clinical improvement in the headache, even after two years of refractory pain to other treatments.

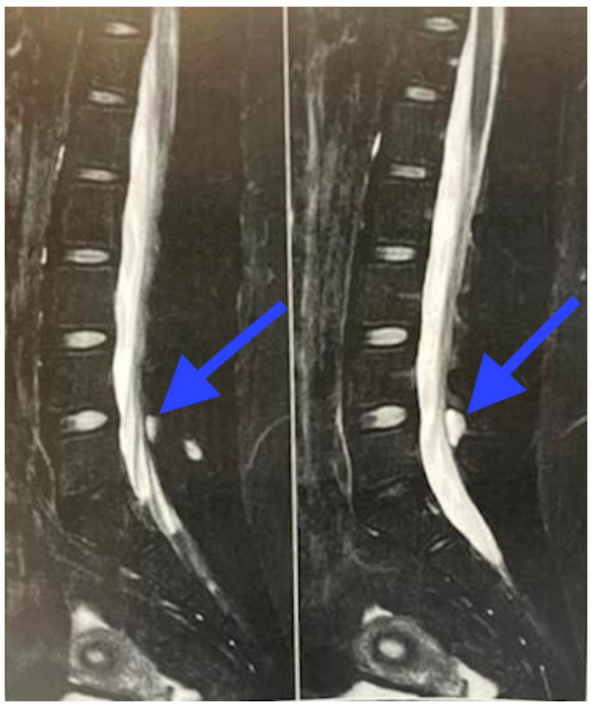

Figure 1. Magnetic resonance imaging (MRI) demonstrating a cerebrospinal fluid fistula, at lumbar level, L4-L5 (blue arrow).

Discussion

The reported case is rare and challenging to diagnose. A clear epidemiological trend indicates a higher incidence of SIH in female patients.[6] SIH is a rare secondary headache that is difficult to diagnose due to its variable

presentation.[1,2] It is caused by CSF leakage, resulting in hypotension and debilitating headaches.[1,2] Venous cerebrospinal fluid fistulas (VCSF) involve a connection between the spinal subarachnoid space and an epidural spinal vein, allowing CSF loss into the circulatory system, believed to be present in up to 25% of patients with SIH.[1,2]

In the present case, this did not occur. The difficulty in initially associating intracranial hypotension resulting from CSF leakage as the cause of painful complaints was highlighted. This differential diagnosis took shape in the presence of the main complaint of orthostatic headache, followed by compatible neuroimaging findings. Early diagnosis of SIH positively impacts prognosis. In addition to postural headache, clinical manifestations of intracranial hypertension also include neck pain, nausea, cranial nerve palsy, photophobia, hearing changes, and other symptoms.[1,2] Furthermore, this headache is precipitated or exacerbated by Valsalva or cough, present in 88% of patients.[7]

According to the International Classification of Headache Disorders, diagnostic criteria for SIH include CSF pressure <6 cm H_2O, imaging evidence of CSF leakage, headache related to low CSF pressure/leakage, or headache not well explained by other causes.[7] However, diagnosing SIH can be challenging since, in most cases, there is no focal extradural CSF collection indicating the presence of leakage in conventional imaging such as MRI and computed tomography (CT).[7]

MRI typically reveals diffuse meningeal thickening with enhancement, believed to occur due to dural vasodilation and increased contrast concentration in the dural microvasculature and interstitial fluid based on the Monroe-Kellie doctrine. This demonstrates that reduced CSF volume leads to compensatory vasodilation in the brain and meninges. Spinal MRI may show an accumulation of epidural fluid, spinal hygroma, dilated cervical epidural veins, and collapsed dural sacs.[2] Thus, additional tests such as dynamic CT myelography (dCTM) are required for diagnostic confirmation, as it can diagnose CSF fistulas based on a hyperdense paravertebral signal suggestive of the fistula's presence.[7,8]

Although the lack of definitive evidence regarding the best treatment, therapeutic options for SIH include percutaneous glue injection, surgical exploration with ligation, transvenous embolization, epidural blood patching, and conservative measures such as caffeine use, hydration, and bed rest.[7] Among these therapeutic possibilities, although epidural blood patching is reported in the literature as a good approach, resolving symptoms in 64% of cases[9], the surgical intervention adopted in the present clinical case proved to be effective, as the patient reported significant improvement in the debilitating headaches that had been affecting her for the past 2 years.[10]

In a case series of 44 patients, with VCSF fistula initially, 90% underwent blood patching, and only one patient responded effectively to the treatment. Due to the failure of initial treatment, 42 patients required surgical therapy, which showed good effectiveness, with 75.6% experiencing symptom improvement, including approximately 50% achieving complete headache relief, indicating that although blood patching is recurrent, it does not provide lasting symptom relief.[7]

Transvenous embolization is also considered an effective treatment VCSF fistula. In a series of cases, 90% of patients treated with this method experienced headache improvement associated with improved MRI findings of the brain.[3] Moreover, embolization is considered a safe treatment, with no evidence of severe hemorrhagic complications or ischemia and the possibility for all patients to be treated on an outpatient basis.[3]

Conflict interest: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Author's contributions

LSBJ, Conceptualization, Validation, Formal analysis, Review, Resources, Project administration, Supervision; ALM, Conceptualization, Methodology, Validation, Formal analysis, Investigation, Resources, Writing-Original Draft, Project administration, Supervision; AOL, Conceptualization, Methodology, Validation, Formal analysis, Investigation, Resources, Writing-Original Draft, Project administration, Supervision; DCP, Conceptualization, Methodology, Validation, Formal analysis, Investigation, Resources, Writing-Original Draft, Project administration, Supervision; MEBM, Conceptualization, Methodology, Validation, Formal analysis, Investigation, Resources, Writing-Original Draft, Project administration, Supervision; CCSL, Conceptualization, Validation, Formal analysis, Review, Resources, Project administration, Supervision; VEP, Conceptualization, Validation, Formal analysis, Review, Resources, Project administration, Supervision.

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