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CASE REPORT

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수막류와 관련된 두개내압 저하

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Intracranial Hypotension Associated with Meningocele

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Intracranial hypotension (IH), often resulting from a cerebrospinal fluid (CSF) leak, is a notable cause of secondary headaches. Diagnosing IH through clinical assessment and neuroimaging can be challenging. Orthostatic headache (OH), characterized by neck stiffness, nausea, dizziness, phonophobia, and photophobia, is a key symptom of CSF leakage, which may stem from a variety of causes, including falling, surgery, CSF studies, or arachnoid diverticula. This study presents two OH patients with IH who were incidentally found to have an underlying meningocele.
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Key Words: Intracranial hypotension, Headache, Meningocele

Cerebrospinal fluid (CSF) leakage is a common cause of intracranial hypotension (IH), characterized by the onset or worsening of an orthostatic headache,[1] often accompanied by nausea, dizziness, hearing impairment, and tinnitus, with improvement in the supine position.[2] While CSF leaks are usually spontaneous, they can also result from trauma, surgery, or dural puncture.[3] Common treatments include an epidural blood patch or dural tear repair.[4] However, in cases involving congenital anomalies such as meningocele, addressing the underlying anomaly may be necessary.[5] We report two patients with IH and meningocele who were managed conservatively and surgically, respectively.

CASES

1. Case 1

A 27-year-old woman was presented to the emergency department with headaches, nausea, and dizziness. She had no notable medical or surgical history but reported the onset of headaches following a fall on ice. She had had no past history of headaches. Initial symptom relief was achieved with analgesics, and she was discharged. However, symptoms worsened a few days later. Upon admission, the neurological examination was normal, with an orthostatic headache pattern that was triggered by standing up and improved on lying down. The headache affected the entire head, particularly the forehead,

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and persisted throughout the day. It worsened within a minute of sitting or standing, reaching a 7-8 on the visual analog scale (VAS). Relief was immediate upon lying down, being reduced to a 2-3 on the VAS. The headache was bilateral, pulsating, and accompanied by nausea, vomiting, photophobia, phonophobia, and neck stiffness during the headache, but no tinnitus or hearing impairment. Her blood pressure was 130/70 mmHg. Blood and urine tests were normal. Brain magnetic resonance imaging (MRI) was normal (Fig. 1-A), but a cervical-thoracic-lumbar-level (C-T-L) magnetic resonance (MR) myelogram showed a congenital cystic lesion at the level of sacrum 3 to sacrum 4, indicative of a meningocele (Fig. 1-B, C). The axial image of the lumbar spine myelogram showed a sacral dural CSF leak (Fig. 1-C). Initially, epidural blood patches (EBPs) were planned, but the anesthesiologist recommended implementing conservative care first and considering EBPs afterward.[6,7] Moreover, conservative measures such as bed rest, aggressive hydration, and analgesics had already proven effective for this patient.[8] By day 5, her symptoms, including severe headaches, had subsided. She was discharged without having further headaches, having scheduled follow-ups with a neurosurgeon. One month later, however, at a neurosurgeon's outpatient appointment, she mentioned experiencing a mild, tolerable buttock pain that did not require analgesics. It was decided that a follow-up MRI of the pelvic bone be scheduled, along with computed tomography scans of the pelvic bone and lumbar spine.

2. Case 2

A 52-year-old woman was presented with headaches, dizziness, and nausea following a fall on ice. She had a history of hypertension and an ovarian cystectomy. Her past headache history revealed that she had menstrual migraine occurring 1-2 days per month. A neurological examination on admission via the emergency department was normal. Her blood pressure was 135/90 mmHg. She was taking hypertension medicines such as amlodipine (5 mg/day), olmesartan (40 mg/day), and hydrochlorothiazide (12.5 mg/day). Blood and urine tests

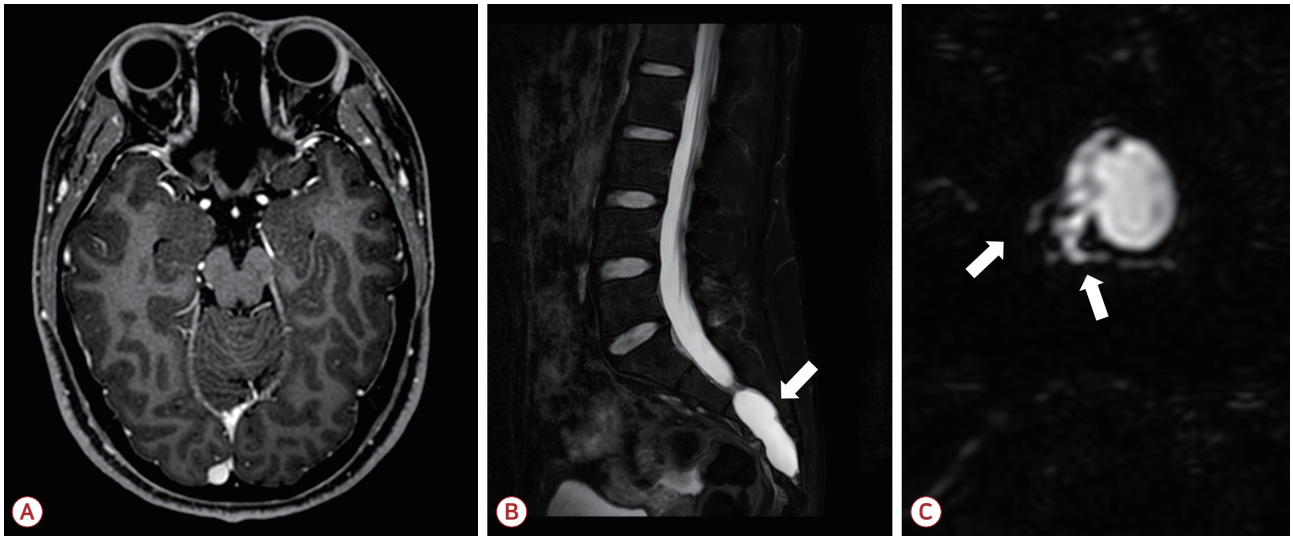

Figure 1. MRI of a 27-year-old woman. (A) 3D T1-weighted images showing a normal brain. (B) T2-weighted sagittal lumbar spine non-contrast myelogram illustrating an underlying intrasacral meningeal cyst (white arrow). (C) Maximum intensity projection reconstruction axial image of lumbar spine non-contrast myelogram showing sacral dural leak (white arrows). MRI; magnetic resonance imaging.

were normal. She had an orthostatic headache, and the headaches were accompanied by nausea, vomiting, photophobia, neck stiffness, and tinnitus. The throbbing and pounding headache affected the entire head without a specific focal point and persisted throughout the day. It intensified within a minute of sitting or standing,

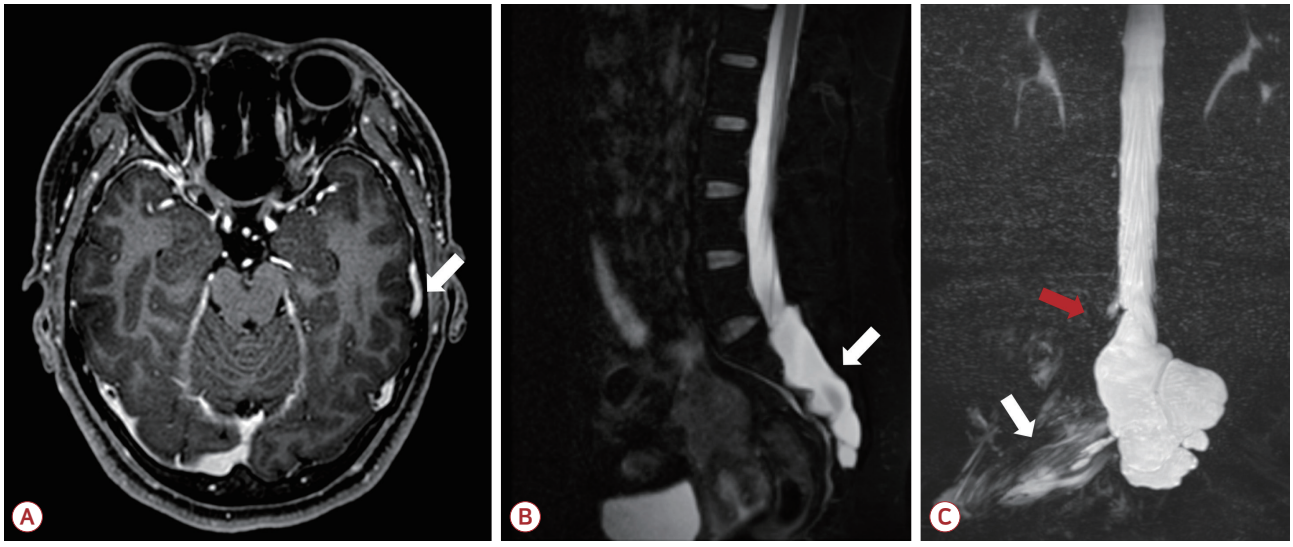

Figure 2. (A) The MRI findings of a 52-year-old woman indicate a small subdural effusion in the left parietal convexity (white arrow), as shown in the fluid attenuated inversion recovery image. (B) T2-weighted sagittal images of the lumbar spine non-contrast myelogram display an underlying intrasacral meningeal cyst (white arrow). (C) The sagittal maximum intensity projection reconstruction image of the lumbar spine non-contrast myelogram demonstrates sacral dural leak (white arrow) and extradural cerebrospinal fluid along the right piriformis muscle (red arrow). MRI; magnetic resonance imaging.

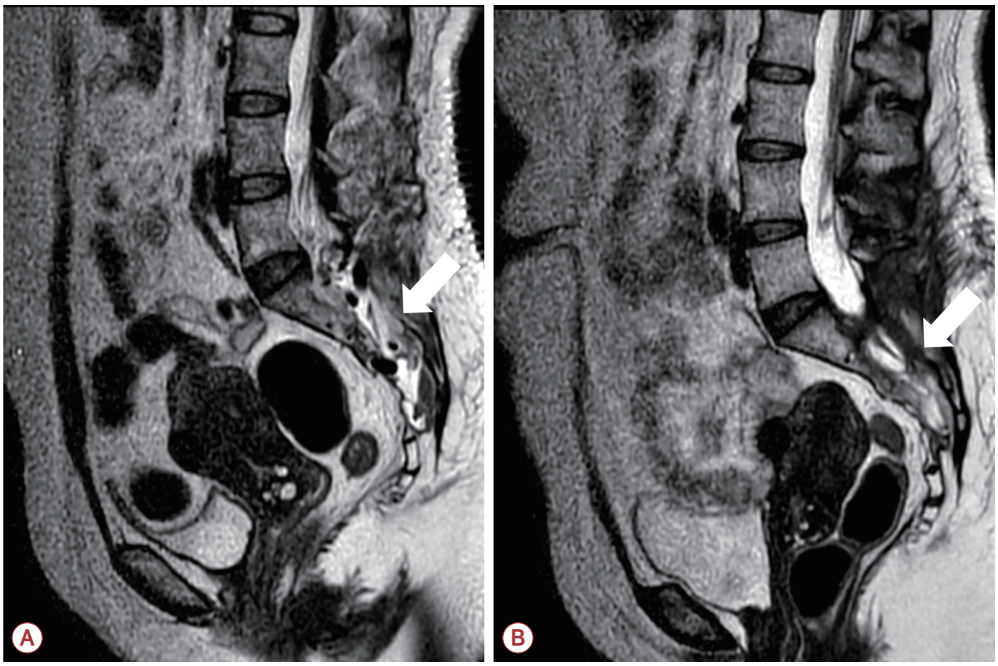

Figure 3. (A) Post-operative T2 sagittal images of the non-contrast lumbar spine MRI of a 52-year-old woman, taken three days after surgery, demonstrate complete removal of the meningeal cyst (white arrow). (B) The follow-up MRI taken 4 months after surgery revealed no recurrence of the meningeal cyst (white arrow). MRI; magnetic resonance imaging.

reaching a 5-6 on the VAS. Immediate relief was experienced upon lying down, reducing the pain to a 0-2 on the VAS. Brain MRI revealed a small subdural effusion in the left parietal convexity (Fig. 2-A). The C-T-L MR myelogram showed not only sacral dural leak and extradural CSF along the right piriformis muscle suspected to originate from the right side S2 and S3 levels but also underlying intrasacral meningeal cyst and ductal ectasia (Fig. 2-B, C). A congenital sacral meningocele was identified, and initially, non-invasive conservative treatment was planned. After 5 days of unsuccessful conservative treatment, surgical removal was recommended during the neurosurgery consultation due to the diagnosis of Tarlov cysts at sacral levels 1 to 4 (Fig. 2-B, C). Subsequently, on the seventh day of hospitalization, the patient underwent a surgery involving the removal of an arachnoid cyst at the lumbo-sacral level and the repair of an arachnoid and dura tear with a muscle flap. A post-operative C-T-L spine MRI was conducted 3 days later (Fig. 3-A). Following the procedure, the patient's headaches resolved, and she was discharged. During one-month of outpatient follow-up, she reported no headaches or neurological abnormalities. A follow-up C-T-L spine MRI performed four months post-surgery revealed no evidence of recurrence and no unusual post-operative change (Fig. 3-B), obviating the need for further follow-up.

DISCUSSION

Our patients can be diagnosed using the International Classification of Headache Disorders, 3rd edition (ICHD-3), under code 7.2.3, headache attributed to spontaneous intracranial hypotension (SIH). The diagnostic criteria for 7.2.3 are as follows: 1) The headache fulfills the criteria for 7.2, headache due to low CSF pressure, and criterion C below. 2) Absence of a procedure or trauma known to be able to cause CSF leakage. 3) Headache has developed in temporal relation to the occurrence of low CSF pressure or CSF leakage, or has led to its discovery. 4) Not better accounted for by another ICHD-3 diagnosis. The meningocele observed in our patients is thought to be pre-existent, and all patients developed orthostatic headaches following a fall, which is a common occurrence in IH. Imaging studies confirmed CSF leakage, and no special procedures, such as a dural puncture, were performed. Thus, the diagnostic criteria for headache attributed to SIH as outlined in ICHD-3 were met.

The treatment of headache attributed to SIH typically begins with a conservative one for up to 2 weeks. Subsequently, an EBP may be considered a therapeutic option.[9] Lützen et al.[10] found that sacral dural leaks accounted for SIH in nine out of 149 patients (6%). All patients with a sacral leak in the cohort were women, similar to the findings in this case report. In contrast to this study, however, eight out of nine patients with confirmed sacral leaks received at least one EBP at the lower sacrum in the reversed Trendelenburg position. In the study by Lützen et al.,[10] all patients underwent a clinical follow-up; six out of nine were monitored for more than 3 months post-treatment (ranging from 3 to 18 months). However, three out of nine patients had clinical follow-up for less than 3 months. A favorable clinical outcome was observed in six out of nine patients, with symptoms significantly improved or completely resolved. Two patients experienced mild to moderate improvement, while one patient showed no clinical improvement to date. Like this study, the study by Lützen et al.[10] also highlighted sacral dural tears as a potential cause of SIH. In contrast to the treatment approaches discussed here, which include conservative management or surgery, the study by Lützen et al.[10] advocates for EBP

in the reverse Trendelenburg position as a therapeutic approach.

This study emphasizes the connection between meningocele and orthostatic headaches precipitated by falling. Recognizing underlying structural abnormalities, such as those associated with meningocele, is essential. A comprehensive evaluation, including imaging studies, facilitates effective assessment and management of the condition.

The findings from this case study underscore the relevance of identifying structural abnormalities like meningocele in the management of falling-induced orthostatic headaches. Further research is warranted to explore the efficacy of treatments such as epidural blood patch in cases involving sacral meningocele, as highlighted by the outcomes observed in the above-mentioned report on the topic.

This study has several limitations. First, it relies on data from a single medical center with a relatively short-term follow-up period, which restricts the evaluation of long-term outcomes. Additionally, the study includes only two cases of IH, which limits the ability to draw broad or definitive conclusions. Second, while the study reports an improvement in IH associated with a large meningocele, it lacks follow-up neuroimaging to confirm this improvement. Successful treatment should be assessed not only by symptomatic relief but also by complete resolution of the CSF leak, as evidenced by imaging.

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