




Spontaneous Intracranial Hypotension Due to CSF Leak at Multiple Spinal Levels—A Case Report and Literature Review

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Abstract

Introduction Spontaneous intracranial hypotension (SIH) is a misdiagnosed condition characterized by low cerebrospinal fluid (CSF) volume and an orthostatic headache that worsens with upright posture due to CSF leak at spinal dural defects. SIH due to CSF leak at multiple spinal levels is a rare condition, and only a few cases have been reported in the literature. Here, we report a case of SIH with a CSF leak at multiple spinal levels. The treatment options and efficacy of epidural blood patch (EBP) injection were discussed in this article.

Case Report A 36-year-old man presented with complaints of orthostatic headache for 2 months and a computed tomography myelogram demonstrated CSF leaks at multiple spinal levels through the dural defects at C5-C6, C6-C7, C7-T1, T1-T2 levels, and at L1, L2, L3, and L4 vertebral levels on the left side. He was managed with a targeted EBP injection and he improved symptomatically within 24 hours of the EBP injection.

Keywords

- ▶ spontaneous intracranial hypotension
- ▶ multiple spinal CSF leaks
- ▶ epidural blood patch injection

Summary SIH is an entity that results from CSF leakage, often through spinal dural defects due to mechanical tear by osteophyte complex, disc prolapse, connective tissue disorders, CSF-venous fistula, and leaking meningeal diverticula. SIH can result from single or multiple dural leaks, but spinal leaks at multiple levels are uncommon. To the best of our knowledge, a total number of 163 cases have been reported in the literature so far. With good success rates, targeted EBP injection with autologous blood is an effective treatment option for both single-level and multiple-level spinal leaks.

Introduction

Spontaneous intracranial hypotension (SIH) is a commonly misdiagnosed condition characterized by low cerebrospinal fluid (CSF) volume and orthostatic headache that worsens with upright posture due to CSF leak at spinal dural defects.

The International Classification of Headache Disorders, 3rd edition, lists the diagnostic criteria for headache due to SIH.¹ It can occur at any age, but most of the patients are middle-aged and there is a female preponderance with a female to male ratio of 1.5:1.² It can occur at a single site or multiple sites, although CSF leaks occurring at multiple spinal levels

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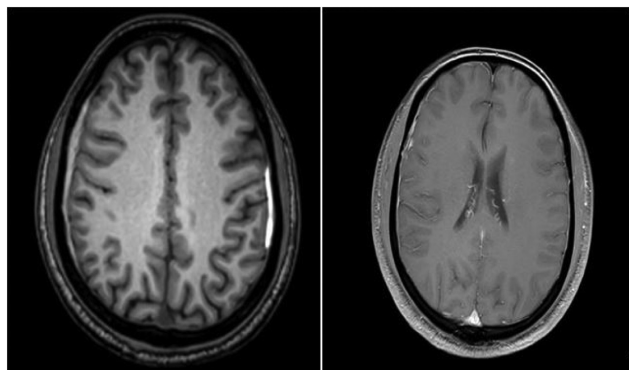


Fig. 1 T1-weighted image showing bilateral convexal subdural hemorrhage of different ages and on contrast admission, meningeal enhancement was noted.

are rarely reported.³ A total number of 163 patients with multiple spinal leaks have been reported in the literature so far.^{3,4} Here, we report a case of SIH with CSF leak at multiple spinal levels, occurring at cervical, thoracic, and lumbar levels. The treatment options and efficacy of targeted epidural blood patch (EBP) injection were discussed in this article.

Case Report

A 36-year-old man with no known comorbidity presented with complaints of an orthostatic headache that worsened in upright posture, associated with multiple episodes of vomiting for 2 months. A clinical examination showed no neurological deficit. His magnetic resonance imaging (MRI) brain showed bilateral convexal subdural collections in the right frontoparietal and left posterior parietal regions with diffuse pachymeningeal thickening and sagging of the brain (**Fig. 1**), features suggestive of intracranial hypotension. However, MRI myelography showed no evidence of any CSF leak. A computed tomography (CT) myelogram was performed with 10 mL iohexol that demonstrated spinal CSF leaks at multiple levels through the dural defects at C5-C6, C6-C7, C7-T1, and T1-T2 levels (**Fig. 2**) on either side, tracking both ventrally and dorsally along the course of exiting nerve roots and through the dural defects at L1, L2, L3, and L4 vertebral levels (**Fig. 2**) on the left side. He was managed with targeted EBP injections with 6 and 20 mL of the patient's own blood into the C7-T1 and L2-L3 epidural spaces, respectively, under fluoroscopic guidance (**Fig. 3**). His headache was relieved within 24 hours of the EBP injection. At 3 weeks' follow-up, the patient had complete resolution of symptoms.

Discussion

SIH is an entity that results from CSF leakage, often through spinal dural defects due to mechanical tear by osteophyte complex, disc prolapse, connective tissue disorders, CSF-venous fistula, and leaking meningeal diverticula.^{2,5} SIH often presents with headaches that are orthostatic but not always, visual disturbances, nausea, vomiting, neck pain, and

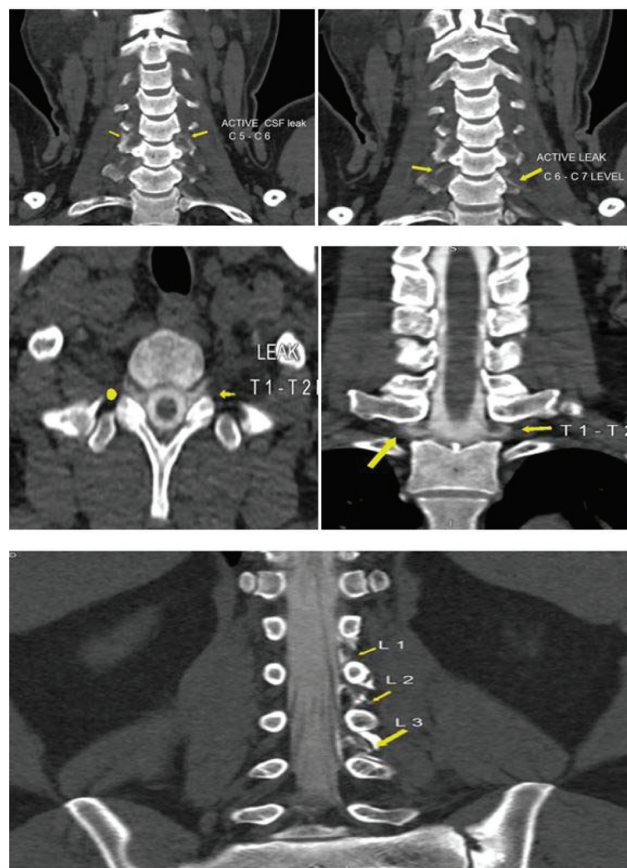


Fig. 2 Computed tomography myelogram showing extravasation of contrast at multiple spinal levels as marked.

other manifestations due to stretching of cranial nerves and the brainstem.⁶ The CSF leak leads to intracranial CSF volume depletion resulting in stretching of the bridging veins, leading to subdural collections and compensation of CSF volume by an increase in intracranial venous blood, which causes engorgement of venous sinuses and pachymeningeal thickening on MRI.⁶ SIH is identified clinically most often, although MRI myelogram or CT myelogram or radionuclide cisternography is required to identify the location of the spinal leak.^{5,7} SIH can result from single or multiple dural leaks, but multiple spinal leaks are uncommon.^{3,8,9} Bhoi et al reported a case of SIH in a 19-year-old girl patient due to recurrent spinal leaks at multiple levels involving lower thoracic and lumbar levels.⁸ In a case series by Schievink et al, SIH due to multiple CSF leaks was seen in four patients with lateral leaks, none of the patients with ventral leaks, and in nine patients with CSF-venous fistulas.³ A meta-analysis by D'Antona et al reported that 24% of the cases had multiple spinal leaks, but the number of cases included in the study was not clear.¹⁰ In a case series by Upadhyaya and Ailani, a total of 149 patients had CSF leaks at more than one site. The most common sites are the cervicothoracic junction and the cervical region.^{3,4,10} In our case, the CSF leak was noted at all levels, including cervical, thoracic, and lumbar levels (**Fig. 2**). The leak was observed bilaterally and in dorsal and ventral locations at the cervical and thoracic levels,

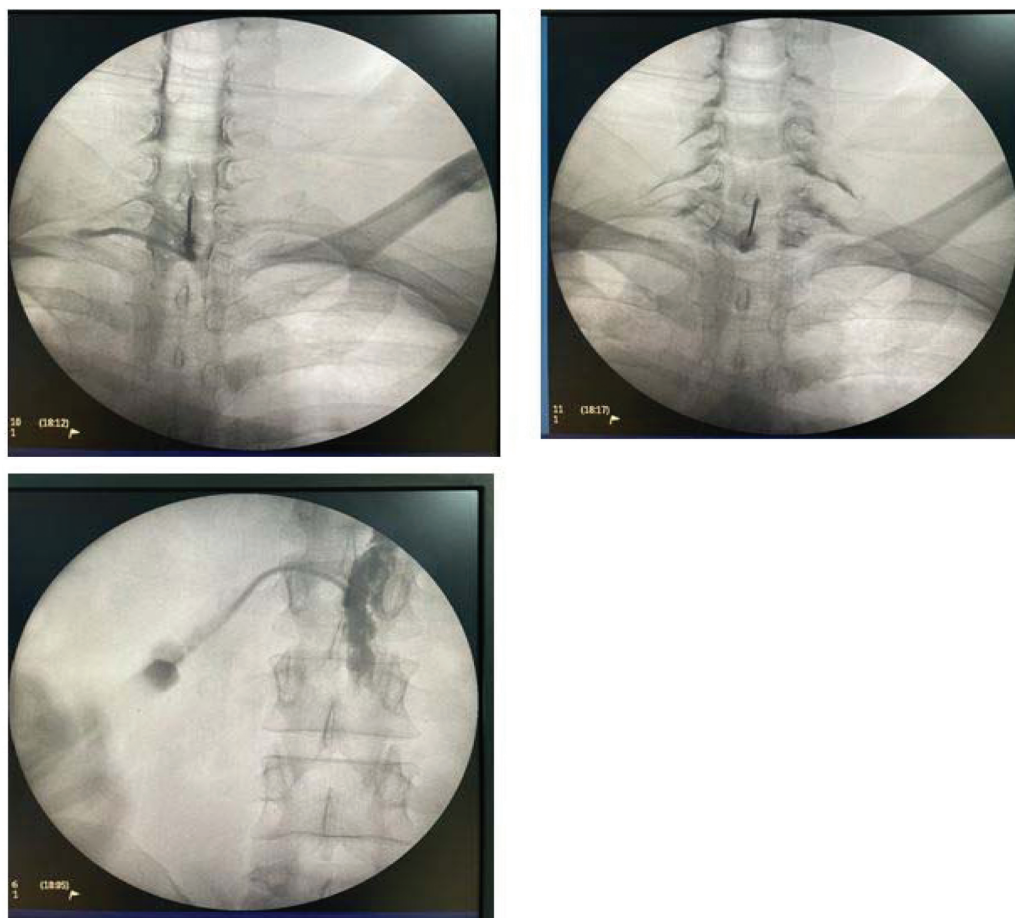


Fig. 3 Epidural blood patch injection under fluoroscopic guidance.

whereas the leak was noted on the left side at the lumbar levels.

The treatment options include rest, analgesics, caffeinated drinks, hydration, abdominal binder, EBP injection, fibrin glue injection, and surgical repair.¹¹ Conservative management for SIH can be attempted at first, but success rates have been reported to be low.^{4,5} A targeted EBP injection is an effective treatment for single or multiple site spinal leaks that involves injecting a small volume of autologous blood into the epidural space under fluoroscopic guidance, which relieves 80 to 90% of intracranial hypotension headaches.⁵

The exact mechanism of EBP injection is not known. It acts by causing a tamponade effect over the dural defect. Later, the fibrin products cause fibroblastic activity and collagen formation, resulting in scar formation and occlusion of the defect completely.⁵ A targeted EBP injection under fluoroscopic guidance has higher efficacy when compared with a blind EBP injection.⁵ However, there is still controversy in the literature about the efficacy of EBP injection, and no proper guidelines have been laid for the treatment of SIH with multiple spinal CSF leaks. Several studies reported low success rates due to failures in identifying the exact location of the CSF leak and blind injection of EBP.^{5,12} CT myelograms and Tc99 DTPA scans are required to determine the precise location and number of CSF leaks.^{4,10} Spinal leaks that occur ventrally cannot be effectively managed by EBP

injection via an interlaminar approach because they cause compression dorsally, resulting in low success rates. CSF leaks occurring ventrally and through nerve roots are well managed by the transforaminal approach. Approximately 20 mL of autologous blood is recommended for EBP injection through an interlaminar approach and 5 mL for a translaminar approach and higher volume does not add any clear beneficial outcomes but rather increases the risk of complications like infection and back pain.⁵ However, a meta-analysis by D'Antona et al reported that a larger volume of blood results in higher success rates.¹⁰ EBP can be injected multiple times for refractory cases with an interval period of 5 days to avoid spinal cord compression.⁵ The success rate of EBP injections ranges from 30 to 70%, but the initial EBP is reported with a success rate of 93%, including partial and complete relief of symptoms.^{4,5,13} Thus, EBP injection requires further validation and a proper guideline in the management of SIH with single and multiple spinal leaks.

Conclusion

SIH is a common cause of persistent orthostatic headaches and related symptoms. SIH due to multiple spinal leaks has been rarely reported in the literature and is often misdiagnosed. The diagnosis needs high suspicion from the clinician and a confirmatory cisternogram to identify the site

of CSF leak. With good success rates, EBP injection with autologous blood is an effective treatment option for both single-level and multiple-level spinal leaks.

Conflict of Interests

None declared.

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