



A Rare Presentation of Spontaneous Intracranial Hypotension

Nadir Görülen Spontan İntrakraniyal Hipotansiyon Olgusu

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Abstract

A 55-year-old man presented with coma induced by spontaneous intracranial hypotension (SIH). In magnetic resonance imaging, sagging and restricted diffusion in the splenium of the corpus callosum, diffuse pachymeningeal enhancement, and bilateral subdural hematomas were observed. Cerebrospinal fluid leakage was detected in the thoracic region and the patient was treated with a targeted epidural blood patch. After the treatment, the lesion in the corpus callosum disappeared without any residual abnormality and the patient fully recovered. SIH can be life-threatening and result in various clinical manifestations from mild orthostatic headache to deep coma. Targeted epidural blood patch treatment seems effective and lifesaving. SIH should be considered in the differential diagnosis of transient corpus callosum splenium lesion.

Keywords: Corpus callosum splenium, spontaneous intracranial hypotension, coma, epidural blood patch

Öz

Elli beş yaşında erkek hasta kliniğimize spontan intrakraniyal hipotansiyonun (SİH) yol açtığı koma tablosu ile geldi. Manyetik rezonans görüntülemesinde korpus kallosum spleniumda difüzyon kısıtlaması, diffüz pakimeningeal boyanma ve bilateral subdural hematom görüldü. Torakal bölgede beyin omurilik sıvısı kaçağı saptandı ve hedef bölgeye yönelik epidural kan yaması işlemi uygulandı. Tedavi sonrası korpus kallosum lezyonu ve hastanın klinik durumu tamamen düzeldi. SİH hayatı tehdit edebilen bir hastalıktır. Ortostatik baş ağrısından ciddi komaya kadar değişebilen klinik tablo ile karşımıza çıkabilir. Kaçak bölgesine yönelik yapılan epidural kan yaması etkilidir ve hayat kurtarıcı olabilmektedir. Geçici korpus kallosum splenium lezyonunun ayırıcı tanısında yer almalıdır.

Anahtar Kelimeler: Korpus kallosum splenium, spontan intrakraniyal hipotansiyon, koma, epidural kan yaması

Introduction

Spontaneous intracranial hypotension (SIH) is caused by cerebrospinal fluid (CSF) leakage. Although the most common clinical manifestation is orthostatic headache, SIH can rarely present with coma due to hypoxemia caused by venous engorgement. Therefore, SIH must be considered in the differential diagnosis of stupor and coma.

The presence of transient lesions in the splenium of the corpus callosum (SCC) has been described in various disorders including epilepsy, demyelinating diseases, infections, and hypoglycemia. To our knowledge, no reports have shown an association of SIH with transient SCC lesions. We present a patient with SIH who showed

transient restricted diffusion in SCC, which was complicated with deep coma, who was treated successfully with a targeted epidural blood patch (EBP).

Case Report

A 55-year-old man was admitted to our intensive care unit due to gradually worsening headache and confusion, which he had had for 1 week. He had a 35-year history of hypertension and epilepsy and had been using fenitoin on a regular basis. On admittance, he was conscious, and fulfilled simple orders but not complex ones. The neurologic examination was unremarkable except for dysarthria and truncal ataxia. Routine blood tests were

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normal. Brain computed tomography (CT) revealed bilateral subdural fluid collections (Figure 1). To elucidate the etiology of the clinical manifestations, brain magnetic resonance imaging (MRI) was performed. On diffusion-weighted images, diffusion restriction was seen in the SCC (Figure 2). On T2-weighted images, bilateral subdural fluid collections, hyperintensity in the SCC and narrowing of the ventricles and sulci were observed. Multiplanar postcontrast images showed diffuse pachymeningeal enhancement, sagging of the SCC, narrowing of the suprasellar cisterna, enlargement of the pituitary gland, and distension of the dural venous sinuses (Figure 3). MRI findings were compatible

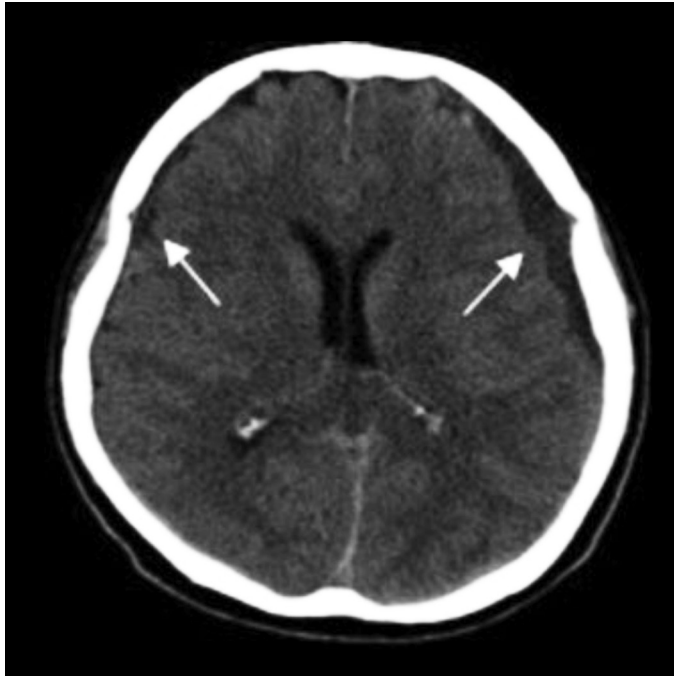


Figure 1. Computed tomography without intravenous contrast injection shows bilateral subdural collections and narrowing of ventricles and sulci

with SIH. During clinical follow-up, the patient revealed a decline in consciousness; he opened his eyes with painful stimulus and localized the pain with both arms. After a couple of hours, he was intubated and mechanically ventilated due to apnea. A second diffusion-weighted MRI showed new lesions with restricted diffusion in the pons and right posterior temporal cortex. We decided to perform an EBP, and to be able to identify the site of CSF leakage, an MR myelography was taken. A small nerve root sleeve cyst was found at the left T7-8 level in MR myelography (Figure 4). We thought that the cyst level was the probable site of leakage because we observed a small amount of fluid signal around the cyst. We decided to perform targeted EBP rather than a regular lumbar EBP because of the high settlement of probable CSF leak.

An epidural catheter was inserted at the T7-8 interlaminar level using the paramedian approach under the fluoroscopic guidance. After confirming epidural placement of the catheter by injecting 5 mL of iodinated contrast agent, 10 mL of autologous blood was injected into the epidural space through the catheter. The day after the blood patch treatment, the patient was extubated and his level of consciousness improved dramatically. A week later, he fully recovered and had a normal neurologic examination. A follow-up MRI on hospital day 10 demonstrated decreased intracranial venous engorgement, decreased subdural hematomas, and normalization of the sizes of the ventricles, cisterna, and sulci (Figure 5). The lesion in the SCC disappeared without any residual abnormality; however, the restricted diffusions in the pons and the right temporal lobe evolved into chronic infarctions.

Discussion

The typical presentation of SIH is orthostatic headache, but the disease can manifest with different signs and symptoms, such as neck stiffness and pain, tinnitus, cranial nerve palsies, phonophobia, venous sinus thrombosis, dementia and coma (1).

According to Monroe-Kellie doctrine, the total volume of intracranial structures such as CSF, blood, and brain parenchyma volume is constant. CSF hypovolemia due to CSF leak is compensated by an increase of blood volume such as subdural

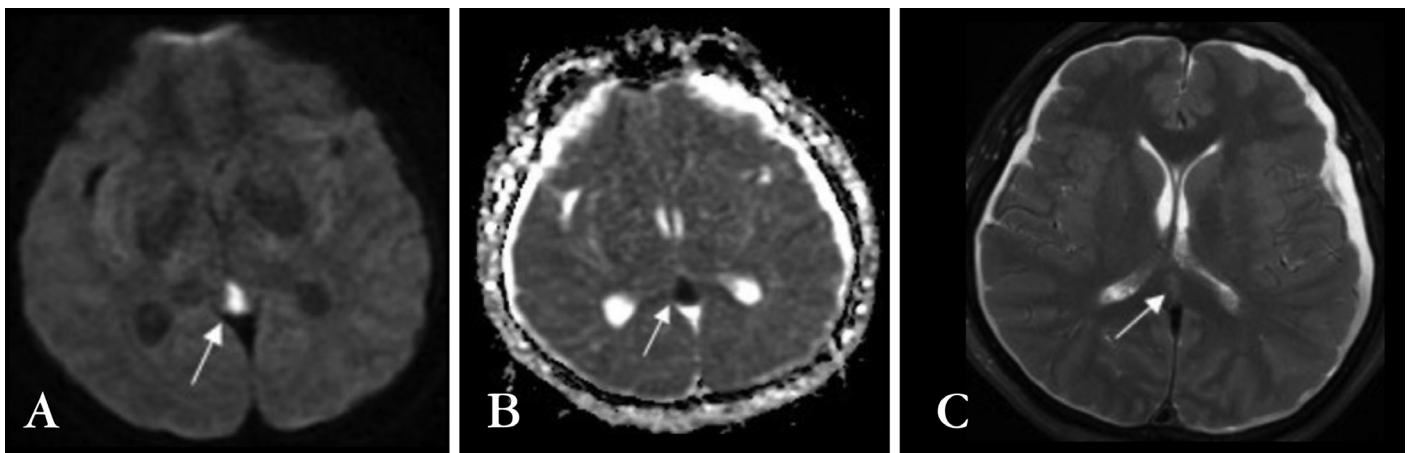


Figure 2. Diffusion and T2-weighted imaging findings in spontaneous intracranial hypotension. A) Diffusion weighted imaging and B) apparent diffusion coefficient map show restricted diffusion in the splenium of the corpus callosum and C) T2-weighted axial, bilateral subdural hematomas, narrowing of ventricles and sulci, and hyperintensity in the splenium of corpus callosum are shown (arrow)

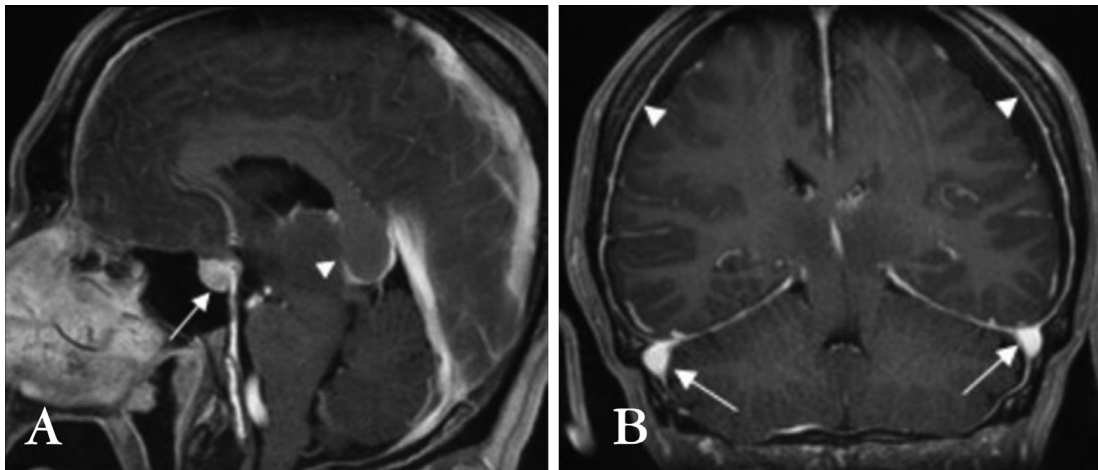


Figure 3. Magnetic resonance imaging findings in spontaneous intracranial hypotension. A) Postcontrast T1- weighted sagittal, B) Postcontrast T1- weighted coronal images. Diffuse pachymeningeal enhancement (arrowheads in B), distended dural venous sinuses (arrows in B), sagging of the splenium of corpus callosum (arrowhead in A), enlargement of the pituitary gland (arrow in A) are also seen



Figure 4. Thoracic magnetic resonance myelography shows nerve root sleeve cysts at T7-8 level on the left side. Fluid signal around the cyst at T7-8 level indicates the probable site of cerebrospinal fluid leakage

hematoma or venous engorgement, and an increase of brain volume as parenchymal swelling. Also, loss of CSF results in reduced buoyancy of the brain, which leads to sagging of brain structures including the diencephalon and brain stem. Sagging of these structures causes a narrowing in the angle between the vein of Galen and straight sinus, which begets a recession in venous drainage of the brain stem. The stagnation in venous flow leads to hypoxemia in the brain stem, which is responsible for the alteration

in the level of consciousness. According to this mechanism, the main underlying cause of coma seems to be venous engorgement rather than direct compression of brain stem (2). Indeed, after the reversal of venous engorgement, the level of consciousness improved in our patient.

Various methods are used for diagnosis such as brain MRI, MR or CT myelography, and radionuclide cisternography. MRI findings are subdural fluid collections, pachymeningeal enhancement, sagging of the brain structures, and engorgement of venous structures (1). As an additional finding, we identified transient hyperintensity in the SCC on diffusion and T2-weighted images.

Transient lesions in the SCC can be seen in some conditions such as encephalomyelitis, epilepsy, antiepileptic drug toxicity/withdrawal, multiple sclerosis, diffuse axonal injury, and hypoglycemic encephalopathy. Despite these different conditions, a common pathophysiologic mechanism that explains splenial predilection in different disorders has not been suggested because of the heterogeneous nature of these diseases (3). On the other hand, a targeted EBP resulted in complete and sustained resolution of the patient's symptoms and diffusion restriction in the SCC. To our knowledge, this is the first case of SIH associated with reversible diffusion restriction in the SCC.

We performed non invasive MR myelography to show the possible site of spinal CSF leakage. In patients with SIH, MR myelography may demonstrate meningeal diverticulae or irregularities along the nerve root sleeves, and epidural fluid collections (4). Although CT myelography is the gold standard method for showing the CSF leakage site, we preferred MR myelography because we did not want to take the risk of increasing CSF hypotension that could have been induced by dural puncture in CT myelography in our comatose patient. Although EBP has traditionally been administered blindly to the lumbar epidural region, increasing reports have shown that effective treatment requires targeted thoracic or cervical blood patches to control CSF leakage (5). The success of targeted EBP in our case may constitute more evidence that a targeted approach should be preferred over blind lumbar intervention when the leakage site rests high in the spine.

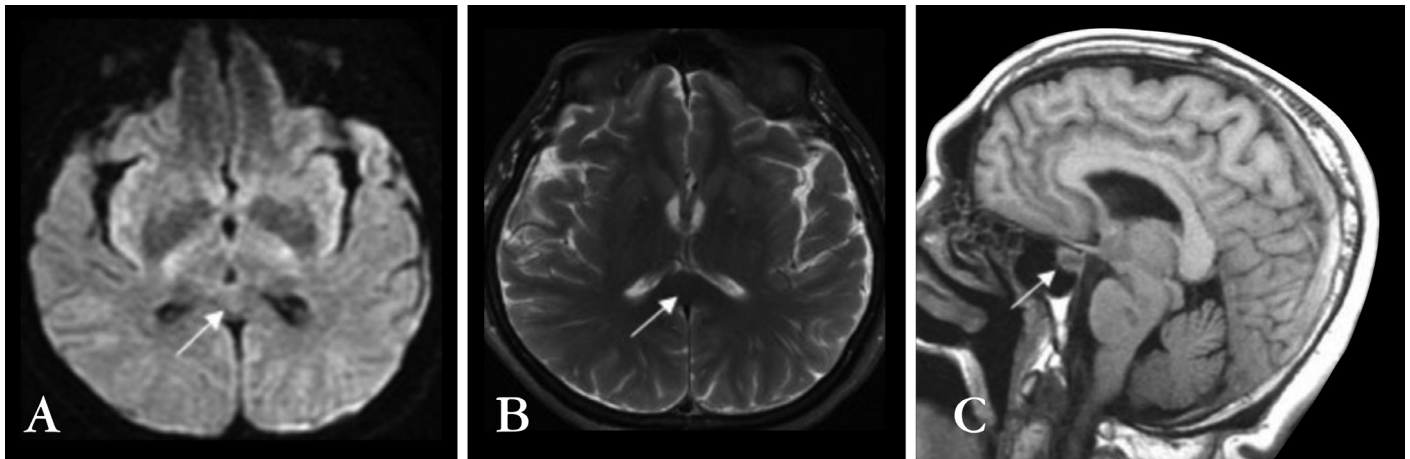


Figure 5. Magnetic resonance imaging findings after targeted blood patch treatment. A) Diffusion weighted imaging, and B) T2 axial images show that the splenial lesion disappeared without any residual abnormality. C) Sagittal T1-weighted image shows normalization of the sizes of the pituitary gland and suprasellar cisterna, and reversal of splenial sagging

In conclusion, SIH presenting with confusion and coma has rarely been reported. A change in headache pattern or consciousness should alert the physician to the possibility of the development of complications, such as subdural hematoma, cerebral venous thrombosis or stroke. In patients for whom initial conservative treatment fails, EBP, preferably delivered at the level of spinal CSF leaks, seems to be effective. SIH should be considered in the differential diagnosis of transient lesions in the SCC.

Ethics

Informed Consent: Consent form was filled out by all participants.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: C.U., V.Ç., Concept: C.U., V.Ç., Ş.A., Y.B., Design: Y.B., Data Collection or Processing: Ş.A., Analysis or Interpretation: V.Ç., Literature Search: C.U., Writing: C.U., Y.B.

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