Sinus Thrombosis in a Patient with Intracranial Hypotension: a Suggested Hypothesis of Venous Stasis

A Case Report

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Key words: intracranial hypotension, cerebrospinal fluid leakage, thrombosis

Summary

We describe the case of a 26-year-old man with orthostatic headache. Cerebral angiography revealed thrombosis in the sagittal sinus. Spine MRI showed cerebrospinal fluid collection at the C1-2 level. We performed blood patch and the symptoms disappeared. We report a rare case of intracranial hypotension caused by CSF leak and describe our hypothesis that SIH can change the velocity of cerebral blood flow and cause thrombosis.

Introduction

Spontaneous intracranial hypotension (SIH) has become a well-understood disease during the last a couple of decades [1-4]. It is generally accepted that SIH is caused by CSF leakage from the dural sac and produce a persistent headache, especially an orthostatic headache. In many cases, subdural fluid collection is common and also combined with meningeal irritation [5-8]. However, little is known about the relationship between intracranial hypotension and cerebral blood flow. There have been few reports regarding thrombus formation in SIH, to our knowledge [9-14]. We experienced the case of a young adult with orthostatic headache who showed evidence of CSF leakage at the C1-2 level and thrombosis at the superior sagittal sinus. We suggest that CSF leakage brings about increased intracranial blood volume and causes the stasis of cerebral blood flow, especially at venous drainage, which leads to thrombosis. The aim of this case report is to ascertain the relationship between intracranial hypotension and thrombus formation.

Case Report

A 26-year-old man who had no remarkable medical or trauma history visited our department. He had neither underlying connective tissue disorders nor risk factors of cerebral venous thrombosis. He had an intractable headache for 1 month, and the headache aggravated when he was in the upright position and completely subsided when he lay flat. The patient underwent brain magnetic resonance (MR) imaging at local clinic, which showed no abnormal findings. He was admitted to a local clinic for three days. After that, his headache became worse and better repeatedly. Finally, his orthostatic headache became intolerable, and he was transferred to our department. He had no neurological deficits except for an orthostatic headache when he initially visited our department. However, brain computed tomography (CT) showed sparse areas of high density along the sylvian cistern on both sides. Thus, we performed diagnostic cerebral angiography to rule out spontaneous subarachnoid hemorrhage and obtained an unusual finding. We also performed a three bottle test of CSF through lumbar puncture. The opening pressure was 50 mmH_2O and the color was clear and did not show hemorrhage in CSF which meant no evidence of subarachnoid hemor-

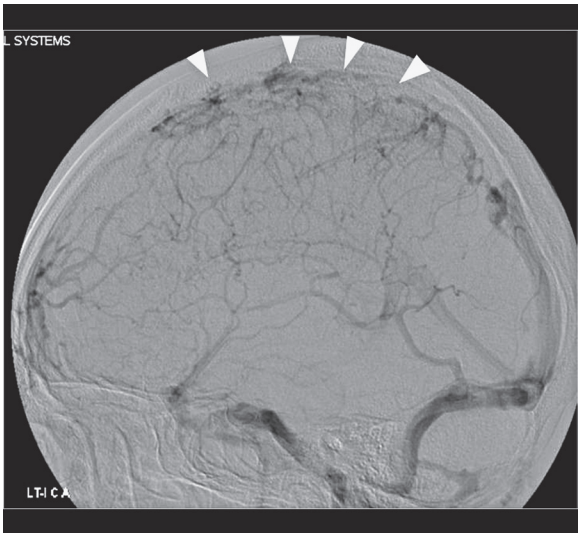

Figure 1 Diagnostic cerebral angiography (left internal carotid artery lateral view) illustrates thrombus formation in the sagittal sinus (arrowheads).

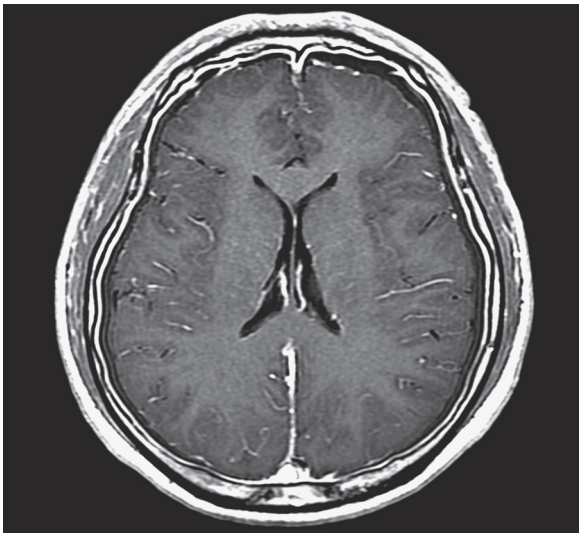

Figure 2 Brain MR imaging shows bilateral subdural effusion and diffuse pachymeningeal enhancement, both of which are typical radiographic findings in patients with spontaneous intracranial hypotension.

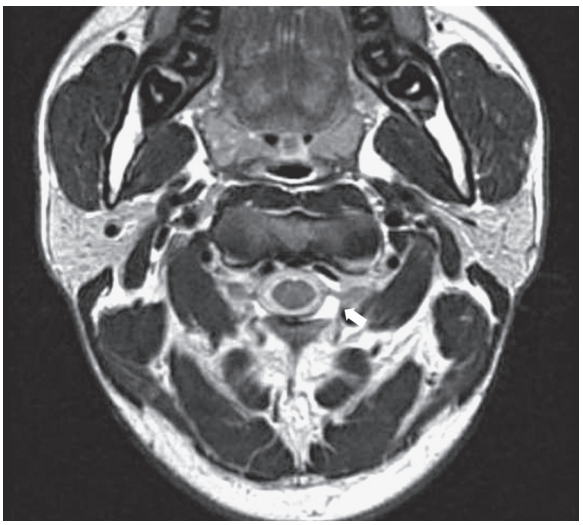

Figure 3 Spine MR imaging demonstrates cerebrospinal fluid accumulation at the C1-2 epidural space on the left side (arrow).

rhage. Thrombotic occlusion at the superior sagittal sinus and stasis of blood flow were found (Figure 1).

Following brain MR imaging performed again two days after cerebral angiography showed bilateral fluid collection at both frontotemporoparietal areas (Figure 2). A thin subdural hygroma was found unlike brain MR imaging that was performed at the local clinic three days before this presentation. A presumptive diagnosis of SIH was made, and spine MR imaging was performed to confirm spontaneous CSF leakage. Spine MR imaging showed CSF collection at the C1-2 level (Figure 3). The patient was definitively diagnosed with SIH due to CSF leakage. An epidural catheter was inserted at the C6-7 level, and blood patch (7 mL) was injected through the catheter. His symptoms were relieved promptly, and no complications occurred. He was discharged from the hospital seven days after the procedure. At the two-week follow-up, the patient had no further headache.

Discussion

In 1938, Renowden et al. first described CSF leakage, decrease of CSF production and increase of CSF absorption as the main causes of SIH [1]. SIH is extremely rare, and there have

been only a few reports of SIH during the last a couple of decades. Cerebral venous thrombosis is also a rare disorder affecting only 0.0005% of the general population, and it occurs in about 2% of patients with SIH [15]. The diagnostic triads of SIH are an orthostatic headache, diffuse pachymeningeal gadolinium enhancement on brain MR imaging and a low CSF opening pressure (below 60 mm of water) [16]. Over 90% of patients have a CSF pressure of less than 60 mm of water (normal value, 65-195 mm of water) [17]. Headache in patients with SIH is diffuse or localized (frontal, temporal or occipital), usually bilateral [18]. It is aggravated gradually and reaches the peak within several hours of onset.

The mechanism of an orthostatic headache is downward displacement of the brain due to loss of CSF buoyancy causes traction on the pain sensitive structure, particularly the dura [19]. SIH exhibits various symptoms including an orthostatic headache, nausea, vomiting, nuchal pain, blurred vision, tinnitus, dizziness, nystagmus, ear fullness and photophobia [21]. In our patient, an orthostatic headache and a nuchal pain were prominent. Previous studies have shown that SIH has a preponderance of females with a male to female ratio between 1:1.75 and 1:4. The mean age of patients with SIH is reported to be between 38 and 42 years, regardless of gender. There is no difference in ethnicity [22-25]. Despite the term 'spontaneous', it is sometimes associated with mechanical stress, which plays an important role in the pathogenesis when the dural sac has a focal, weak point [20,26]. The site of CSF leakage is predominantly at the cervical or thoracic level. SIH is caused by single or multiple spinal CSF leakage, and generalized connective tissue disorder plays a crucial role in the development of spontaneous CSF leakage.

SIH is usually diagnosed by MR imaging [27]. Diffuse meningeal enhancement is a characteristic MR finding. Bilateral or unilateral subdural fluid collection is frequently associated with SIH. CT myelography is the most useful for determining the exact site of CSF leakage [4,28]. However, we did not perform CT myelography because fluid collection was definitely found at the C1-2 level on spine MR imaging. Spine MR imaging is a useful tool for detecting a leak site point indirectly.

Our patient had typical symptoms and brain MR imaging findings. Under a diagnosis of SIH, we performed diagnostic cerebral angiography, which showed thrombosis in the sagittal sinus. The reasons why blood flow stasis occurs have been proposed in the literature [12,15]. Farb et al. [29,30] presented a study about venous distension in SIH patients group by using MR image. They analyzed a diameter of transverse sinus on MR image and showed a venous distension of transverse sinus. Based on the suggestions about cerebral venous thrombosis in patients with SIH, we present the following hypothesis. First, intracranial volume is constant by autoregulation. Second, according to the modified Monro-Kellie hypothesis (intracranial blood volume [CBV] + brain + CSF = constant) [8,31,32], if CSF volume is decreased by leakage, it increases cerebral blood volume. Third, to maintain increased intracranial blood volume, the diameter of the venous structure, such as the sagittal or transverse sinus, increases [12]. Fourth, blood flow velocity is decreased in intracranial blood vessels, especially in the venous phase [12,15]. Finally, repetition of these events contributes to thrombus formation in the venous structure. According to the hydrodynamic theory (continuity equation $S1V1=S2V2$ where S is cross section and V is flow velocity) implying that flow velocity is inversely proportional to cross section, blood flow velocity in the sagittal sinus is slow compared to other vessels because the sagittal sinus has the biggest diameter among intracranial vessels [33]. Furthermore, CSF loss reduces the absorption of CSF into the sinus, and it increases blood viscosity [15].

Surgical repair of the leak site is the standard treatment option; however, since it is not available in most cases, the procedure for sealing the hole of the leak site is the second choice of treatment [3]. It has been demonstrated that autologous epidural patching is the treatment of choice. Fibrin sealant injection is the next choice. Our patient underwent autologous blood patch (7 mL) once, and symptoms disappeared promptly. However, most patients require two or more epidural blood patches. Supportive care, such as hydration, bed rest and caffeine administration, provides only a temporary relief of symptoms [18].

Conclusions

Although a few studies on SIH have been published during the last several decades, these studies have reported not on blood stasis but mainly on treatment in patients with SIH. In this study, diagnostic cerebral angiography showed thrombosis in the sagittal sinus. We put

forward the hypothesis that CSF leakage results in increasing intracranial blood volume and causes the stasis of cerebral blood flow, which leads to venous thrombosis.

Acknowledgment

This work was supported by the Dankook University Research Fund of 2006.

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