Spontaneous Intracranial Hypotension: Diagnosis and Treatment of 2 Cases

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Summary

Intracranial hypotension is a clinical syndrome of decrease in volume or pressure of cerebrospinal fluid (CSF), presenting with orthostatic headache. It is diagnosed by typical clinical presentation and documentation of decrease in CSF pressure, along with the radiological findings. The first treatment option is conservative; bed rest, fluid replacement, caffeine, theophylline, abdominal bandage. If pain persists, epidural blood patch is considered the gold standard treatment. In this article patients resistant to medical treatment were treated with epidural blood patch for different etiologies; also diagnosis, radiological features and treatment approaches were discussed.

Key words: Intracranial hypotension, epidural blood patch, headache

INTRODUCTION

Intracranial hypotension is generally characterized by orthostatic headache. Orthostatic headache gets worse when the patients stand up. The headache increases with laughing, coughing, jugular venous compression, Valsalva manoeuvre and it is resistant against analgesic treatment. And also there would be nausea, vomiting, neck pain, vertigo, horizontal diplopia, hearing disorders, tinnitus, pins on face and upper extremity radicular symptoms. International headache society (IHS) evaluated intracranial illnesses that were not vascular in The International Classification of Headache Disorders (ICHD-III) in 2013. The etiologic factors are post lumbar puncture, post spinal anesthesia, or spinal injury due to dural rupture with CSF leakage, general dehydration, uremia, diabetic coma, metabolic abnormalities causing decreased
production or increased reabsorption of CSF or idiopathic\(^{(9)}\). Classical scanning symptoms due to loss of CSF in subarachnoid distance with increased total intracranial blood pool volume. Dural contrast retention with subdural effusion and prolapse of brain according to the period of CSF leakage and amount are seen before progressive symptoms become clear in venous system. Cranial magnetic resonance imaging (MRI) with contrast is an effective diagnostic method especially on early stage and are diagnosed with brain sag. Although MRI methods is insufficient for showing the place of CSF leakage, computed tomography (CT) or radionuclide sisternography are used with limitations such as, high radiation exposure and low spatial solubility with contrast myelography. Contrast MR myelography is an effective method which doesn't involve radiation to diagnose CSF leakage and has advantages over MRI like spatial resolutions.

Intravenous fluid replacement, caffeine, acetaminophen, abdominal bandage are used for treatment. Blood patch is gold standard for the patients that fail with these treatments\(^{(15)}\). We are going to discuss two cases with different etiology and radiologic features which are resistant to treatment and we treated with blood patch.

**CASE PRESENTATION**

**CASE 1**

A twenty-two year old woman has had shooting headache starting in the frontal area and spreading to the back of the neck for a week. The pain was partially relieved when she lies down, it increases when she sits and stands. MR venous angiography shows prominence of dural venous sinuses with increased inferior convexity of transverse and sigmoid sinuses(Figure 1). The symptoms for intracranial hypotension weren't determined in all of spinal MRI. On Tc-99 DTPA sisternography all body radionuclide screening in place of CSF leakage couldn't be shown when the patient was in a suitable position. LP was done between L3-L4. The patient's CSF pressure was measured as 30 mmH\(_2\)O. In biochemistry of CSF, glucose: 49 mg/dl protein:35 mg/dl simultaneous blood glucose: 85mg/dl were measured. During follow-up relaxation, fluid treatment, caffeine and theophylline didn't relieve the headache. Epidural blood patch were done at L3-L4 and 20 cc autolog blood were administered. The headache was relieved and didn't come back for 6 months.

**CASE 2**

A forty-four year old woman patient came with a headache that started from the neck and spreaded to the left of the face and diplopia. The headache increased when she was sitting up and it decreased while she was lying. Pain was partially relieved by NSAID. Diplopia increased when she looked left and it got better when she closed one of her eyes.

On neurologic examination there was limited lateral gaze of the left. Bilateral fundus examination was normal. Laboratory tests including hemogram, liver and kidney function tests, thyroid function, lipid electrophoresis, erythrocyte sedimentation speed were normal. Infra and supratentorial dural thickening and contrast enhancement were observed at cranial MRI. Displacement of flour of 3.ventricle and sagging of cerebellum and brain stemwere revealed(Figure 2). Cervical MRI was performed and dural contrast leak to extradural space was found betweenC6-C7(Figure 3). The patient was diagnosed spontaneous intracranial hypotension, according to MRI and clinic symptoms and hydration, paracetamol combined with caffeine treatment was done. Theophylline infusion didn't subside the headache. Conservative treatment was not successful, so L3-4 epidural blood patch with 16 ml autologous blood was performed under fluoroscopy. Soon after the headache and diplopia were relieved.
**Figure 1:** A sagittal T1-weighted (A), anteroposterior 3-dimensional TOF MIP (B) and coronal plane TOF (C) MR venous angiography show prominence of dural venous sinuses with increased inferior convexity of transverse and sigmoid sinuses (arrows).

**Figure 2:** Axial susceptibility-weighted (A and B) images show prominence of venous vascular structures. Coranalgadolinium-enhanced T1-weighted image shows enhancement and thickening of dura (arrowheads). Axial T2-weighted (D) and gadolinium-enhanced FLAIR image (E) images show fine subdural collection and dural enhancement (arrows). Sagittal T2-weighted (F) image shows displacement of flour of 3. ventricle and sagging of cerebellum and brain stem.
DISCUSSION

Headache due to intracranial hypotension often resolves spontaneously within two weeks. However, prolonged cases have been reported\(^4\). It is often iatrogenic, traumatic and spontaneous in origin. The etiology of spontaneous CSF leak remains uncertain. Leakage can be for all spinal canal, but often the cervical and thoracic area in orthostatic headache\(^6\). IHS diagnostic criteria of low CSF pressure headache have been identified\(^5\) (Table 1).

Headache starts positionalina few minutes or may be reduced. However, it may take longer in chronic headaches. Headache may be throbbing and sometimes very intense and variable severity feeling of pressure is defined. Headache is usually bilateral\(^{10}\) and usually does not respond to analgesics, often in frontal and occipital areas and, exacerbated by Valsalva maneuver, coughing and laughing. For the diagnosis of spontaneous intracranial hypotension; postural headache, low CSF pressure and relevant MRI findings with clinical evaluation is required\(^3\).

**Table I:** Orthostatic headacheDiagnostic criteria: (3).

| A. Any headache fulfilling criterion C |
| B. Low CSF pressure (<60 mm CSF) and/or evidence of CSF leakage on imaging |
| C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or has led to its discovery |
| D. Not better accounted for ICHD-3 Diagnosis |

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*Figure 3: Axial T2-weighted(A) image shows CSF leak in to the epidural space(arrowheads). Axial T1-weighted MR myelography(B) following intrathecal gadolinium administration shows focal site of CSF leakage consistent with dural defect.*
Typically imaging pattern for intracranial hypotension due to loss of subarachnoid fluid and increased total intracranial blood pooling are: dural (pachymeningeal) contrast enhancement, venous enlargement, pituitary hyperemia, subdural effusion and brain sagging. According to the duration and the amount of CSF leakage, these findings progressive from mild to severe before the venous structures are evident, subdural effusion, dural contrast enhancement and brain sagging may proceed. Contrast enhanced cranial MRI especially in the early stages and uncertain cases is a more effective method compared with CT. However, in 20% of intracranial hypotension cases normal MRI findings are reported.

In some patients, with active CSF leakage, typical clinical history, examination findings and imaging findings may not be displayed. Routine MRI techniques are insufficient to show location of CSF leak. CSF leakage detection techniques used in the CT or radionuclide cisternography but particularly such as high radiation exposure and low spatial resolution are major limitations. Non-contrast and contrast MR myelography in determining CSF leakage does not contain radiation and as high spatial resolution of MRI is provided is an effective method with advantages. Precise MRI findings in patients for diagnose lumbar puncture (LP) to measure the CSF pressure is not required. CSF opening pressure in intracranial hypotension on LP is below 60 mm-H2O. Some patients have been reported in the CSF pressure within normal limits. These patients probably have decreased CSF pressure, but the average remains within normal limits.

In Case 1 is there was a positional headache present. In cranial and all spinal MRI examination findings of intracranial hypotension was not available except intracranial venous congestion at the border. Radionuclide imaging with Tc-99m DTPA cisternography did not show the location of CSF leaks. On top of that LP was performed diagnostically to measure the CSF pressure. The patient's CSF pressure was measured as 30 mm-H2O and headache due to spontaneous intracranial hypotension was diagnosed.

At case 2, the headache was increasing when standing and it was decreasing when lying down. Also diplopia was increasing when she looked left, but it was getting better after closing one of her eyes. Typical radiological findings were for intracranial hypotension on cranial MRI and MR cisternography with contrast to determine CSF leakage. Contrast leakage was followed to extradural area on cervical 6-7 vertebral plane. The headache diagnosis related to hypotension was made before LP wasn't done to measure CSF pressure. After epidural patch for case 2 diplopia regressed. In the literature, patients with CSF leakage from the cervical and thoracic spine performed lumbal epidural blood patch was shown to be effective. However, the epidural blood patch made from the area of the CSF leak chances of success are higher. In our case 2 we chose lumbal region because of the ease of application of lumbal epidural blood patch. If unsucces, we plan to do the regions that have CSF leak. However, in our case we have made a success of lumbal epidural blood patch.

Intracranial hypotension can be treated in most of the patients with conservative approach: bed rest, coffee, hydration, abdominal bandage, analgesics, caffeine, theophylline and corticosteroids. Ergin showed that theophylline infusion was effective on headaches related to intracranial hypotension. If these treatments fail, autolog blood and fibrin glue are patched to leakage area. Epidural blood patch is gold standart treatment for dural leakage. Blood patch of effective for treatment is explained with two basic mechanisms which complete.
each other. Blood patch increases volume of distance, intracranial pressure and stops the leakage by making a buffer\(^{(15)}\). There are kinds of studies about how much blood need to be given. Lung has offered 12-40 ml on lumbar area, 11-20 ml on thoracic area, 6-13 ml of autologous blood in cervico-thoracic junction\(^{(13)}\). The blood that is applied to epidural areas has spread to six cephalic spinal segments and three caudal spinal segments\(^{(14)}\). After first blood patch, rate of success is %36-57\(^{(13)}\). It is recommended to have five days between repeated blood patch. Frequently clinical symptoms regress in 72 hours after this treatment, when it is made from leakage area or upper or down distance. If the level of leakage is known blood can be injected there as targeted epidural blood patch\(^{2}\).

Blood patch is available and a satisfactory treatment when the headache due to intracranial hypotension is unresponsive to other treatment methods. If it is necessary, epidural blood patch can be repeated. Successful outcomes can be increased by clinical and radiologic features.

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