



## Spontaneous Cerebrospinal Hypotension: A Case Report

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### Abstract

Spontaneous cerebrospinal hypotension may be caused by leakage of cerebrospinal fluid. The characteristic symptom is orthostatic headache. MRI scan may yield a number of characteristic abnormalities. Treatment initially is conservative. However, some patients may need a more invasive approach. We present a case of a 40-year-old female with orthostatic headache. In this case there was some delay until adequate treatment was given.

### Keywords

Spontaneous cerebrospinal hypotension, Headache, Lumbar puncture, Liquor hypotension, Epidural blood patch

### Introduction

Post spinal headache after lumbar puncture is a well-known complication. Characteristically the headache occurs when sitting up or standing. The headache will resolve in lying position. Cerebrospinal hypotension however, may also occur spontaneously. We describe the case of a 40-year-old female with spontaneous intracranial hypotension syndrome.

### Case Report

A 40-year-old woman without previous medical history was referred to the neurologist because of serious headache. When working in the garden pulling weeds from the ground she felt a 'pop' in the back of her neck. In the consecutive days she developed a severe headache. The family doctor suspected a tension type headache and prescribed diazepam and naproxen. However, this had no effect. A few days later the patient returned. She experienced a more severe headache while sitting or standing. When she lay down, she had no symptoms. The doctor prescribed a stronger pain medication and referred her to a neurologist.

Based upon the history, the neurologist suggested spontaneous intracranial hypotension. She was treated with bed rest for 72 hours. Unfortunately there were no signs of improvement. Therefore, a MRI of the cerebrum and complete spine with intravenous gadolinium was

ordered. The MRI was performed 5 days after her visit to the neurologist and showed no abnormalities. The diagnosis spontaneous intracranial hypotension syndrome therefore could not be verified and the neurologist decided to do no further interventions. Because of persistent debilitating headaches she repeatedly contacted the neurologist. He eventually decided to perform an epidural blood patch with 15 ml of autologous blood which was injected at the lumbar level L1-L2. At first the headache seemed to resolve. Unfortunately, after just 1 day it returned and the patient was referred to a neurosurgeon.

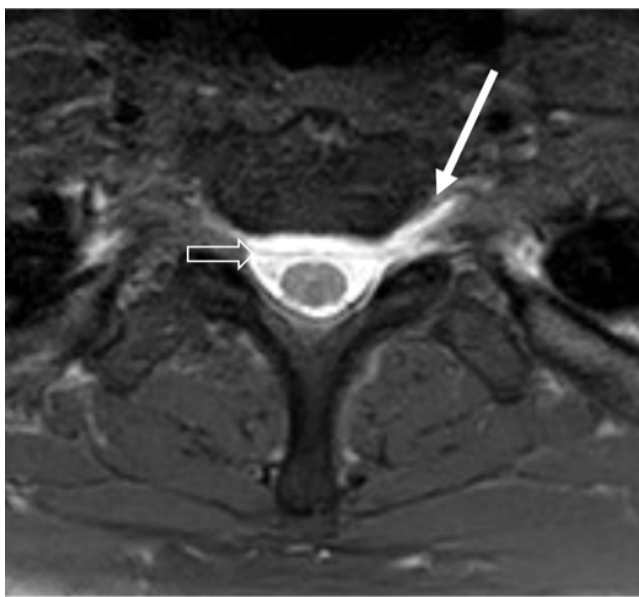
The neurosurgeon ordered a new MRI of the complete spine, only this time gadolinium was given intrathecally. 0.5 ml of undiluted magnevist was injected using a 22 gauge atraumatic needle at the L3-L4 level. Two scans were obtained. The first scan directly after injection of the gadolinium, the second scan 5 hours after the first one. These new scans showed a leakage at the level C7-Th1. It was now decided by the neurosurgeon to consult

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**Figure 1:** Fat suppressed T1 weighted images after intrathecal Gadolinium injection shows cerebrospinal fluid leakage at the level of the first thoracic nerve root on the left side (arrow). The dura can be seen as a fine black line between the opacified intra and extradural space (arrowhead).

an anaesthesiologist-pain specialist, to perform a targeted epidural blood patch under fluoroscopy. The patient was positioned on an operating table in the prone position. The patient underwent the procedure under local anaesthesia in order to monitor any sensory or motor defects. In addition twenty ml of autologous blood was slowly injected in four to five minutes under direct fluoroscopy exactly at the C7-Th1 interspace using an 18 gauge epidural Tuohy needle. During injection care was taken in detecting any increase in injection pressure in order to avoid any risk of spinal cord compression. After this targeted procedure the patient quickly recovered. After a 3 month follow up period the patient had no more complaints. There was no neurological deficit of any kind (Figure 1 and Figure 2).

## Discussion

The leading symptom in spontaneous intracranial hypotension is orthostatic headache. Other symptoms include neck pain, nausea and vomiting. Other, neurological symptoms may also be present. These include cranial nerve palsies, radicular pain of the upper extremities, hearing loss, tinnitus, ataxia, diplopia, epilepsy, altered mental state and death [1-3].

Spontaneous intracranial hypotension predominately occurs in women. The man:woman ratio is 1:2. The mean age is 38-42 years. Underlying connective tissue disease is a predisposing factor. The estimated annual incidence is 5 per 100000 [1-5].

Spontaneous intracranial hypotension syndrome is



**Figure 2:** PA view after placing the needle and injecting contrast.

caused by spinal fluid leakage, resulting in a decreased cerebrospinal fluid volume rather than pressure. As most patients will have a normal cerebrospinal fluid opening pressure [4,6]. Most frequent the rupture is found at the cervicothoracic or thoracolumbar junction [1-3,6]. Leaks can arise from dural weakness involving nerve root sleeves, ventral dural tears caused by disk herniations and cerebrospinal fluid venous fistulas [6].

Under normal circumstances the brain is mainly supported by the cerebrospinal fluid on which it rests. A small part of the brain is supported by pain-sensitive structures such as the meningeal membranes, blood vessels, the cerebral nerves (V, IX and X) and the three upper cervical nerve roots. Decrease in cerebrospinal fluid volume results in traction of the pain-sensitive structures [5]. This mechanism is responsible for some of the described symptoms. The headache therefore predominantly occurs in the upright position. Spontaneous intracranial hypotension headache rarely occurs in the elderly because the mass of the brain decreases with age [3].

In our patient the diagnosis spontaneous intracranial hypotension was not readily recognized. As a result she suffered from debilitating headaches preventing her to perform normal daily activities. Early recognition is important and prevents delay in treatment. The outcome is usually benign but there are several cases in literature with near fatal outcomes [2].

The international Classification of Headache Disorders has identified diagnostic criteria [4,6]. Spontaneous intracranial hypotension is diagnosed based on the patient's history, neurological evaluation, lumbar puncture and imaging studies. An MRI can yield the following abnormalities; meningeal contrast enhancement, downward displacement of the brain and cerebellar tonsils, engorgement of the venous plexus, enlargement of the pituitary, subdural fluid collections and extradural fluid collections. However, in approximately 20% of the cases no abnormalities are found [2,3,5-8]. When initial treatment fails further investigation is advised. Imaging tools include radionuclide cisternography, CT myelography or MRI with intrathecal gadolinium [4,9-11].

Spontaneous intracranial hypotension is initially treated conservatively with analgesics, hydration and bed rest. When conservative therapy fails an epidural blood patch can be performed [3-5,7,12]. In a non-targeted blood patch autologous blood is injected in the lower thoracic or lumbar epidural space. Success rates vary widely. According to Kranz, et al. the success rate range from 30-70%. The success rate increase further with a second blood patch. However this could lead to fibrosis and obliteration of the epidural space. Therefore clinicians should be cautious to administer multiple blood patches [4].

A more successful approach is a targeted blood patch. Autologous blood is injected in the epidural space at the level of the cerebrospinal fluid leak, or 1 level above or below. Reported success rates are 87% [5,6]. A more invasive treatment is surgical repair. This should be considered in patients with an identified leak site and persisting symptoms after 2 or more blood patches [4,5,12].

## Conclusion

Post spinal headache after lumbar puncture is a well-known syndrome. It occurs frequently and is diagnosed easily. However intracranial hypotension may occur spontaneously without obvious trauma. The characteristic symptom is orthostatic headache. Early recognition is important to prevent unnecessary delay in treatment.

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