

SPONTANEOUS INTRACRANIAL HYPOTENSION IN A PATIENT WITH MARFAN'S SYNDROME TREATED WITH EPIDURAL BLOOD PATCH

- A Case Report -

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Summary

Spontaneous intracranial hypotension (SIH) is a well-defined clinical entity that is frequently misdiagnosed. We are reporting a case of 38 years old male who presented with severe headache and an episode of generalized tonic-clonic seizure. He was managed successfully with an epidural blood patch. Understanding of the characteristics, symptomatology, evaluation, treatment options, and prognosis is discussed.

Key words: Epidural blood patch, Spontaneous intracranial hypotension, Marfan's syndrome

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Introduction

Clinical syndrome of spontaneous intracranial hypotension (SIH) characterized by low cerebrospinal fluid (CSF) pressure with no apparent history of dural puncture or trauma was first described by Schaltenbrand in 1938¹. The clinical hallmark of SIH is the presence of orthostatic headache which may be associated with neck pain, nausea, vomiting, diplopia, blurred vision, and distorted hearing².

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SIH results from spontaneous leakage of spinal CSF, presumably due to the weakness in the spinal meninges but the exact cause remains unknown³. In some cases there may be history of a trivial traumatic event. Meningeal diverticula and SIH have been described in several connective tissue disorders such as autosomal dominant polycystic kidney disease⁴, Marfan's syndrome⁵, etc.

We are presenting a case of SIH, relevant diagnostic and therapeutic interventions are discussed.

Case

A 38-yr-old male, known case of Marfan's syndrome, presented in the Emergency Room (ER) with positional headache since 15 days unresponsive to oral analgesic, neck stiffness and an episode of generalized tonic-clonic seizure. His past history included aortic valve and aortic root replacement (for which he was on warfarin) and transient ischemic attack. His examination was unremarkable without any neurological deficit. Routine laboratory investigations were also within normal limits except for prolonged prothrombin time (PT) and International Normalization Ratio (INR), which was 1.56.

Magnetic Resonance Imaging (MRI) of brain and spine showed bilateral small subdural hematomas and cerebral tonsils pointing inferiorly with signs of patchy meningitis. MRI myelogram of the spine showed expansion of thecal sac in lumbosacral region with multiple outpouchings arising at the level of 1st, 2nd and 3rd sacral vertebrae (dural ectasia). Fluid was seen adjacent to posterior paraspinal muscles in the lumbosacral region with no definite leaking tract. In view of the above findings, a diagnosis of spontaneous dural tear leading to CSF leak with SIH was made. Acute pain management service (APMS) was consulted for lumbar epidural blood patch as symptoms were not improving with conservative management. Lumbar epidural blood patch was successfully performed at L4-5 interspace under all aseptic measures using a 16G Tuohy needle (Portex® Epidural minipack Smiths Medical International ASD, Inc. Keene, USA.) with 18ml of autologous blood. Patient remained hemodynamically stable throughout the procedure. Pain was moderately relieved immediately after the procedure and after 48 hours of follow-up there was

complete pain relief without any neurological deficit.

Discussion

Once considered an exceedingly rare disorder, recent evidence suggests that SIH should be considered in the differential diagnosis of new persistent headaches, particularly among young and middle-aged individuals. In an emergency department based study the incidence was found to be five per 100,000⁶. SIH usually results from spontaneous leakage of spinal CSF but the exact cause is unknown. SIH is commonly associated with spinal meningeal weakness secondary to an underlying connective tissue disorder and in about one third of the patients, a history of trivial traumatic event is also present suggesting a role of mechanical factor as well⁷.

The clinical hallmark of intracranial hypotension is the presence of orthostatic headache which occurs or worsens in less than 15 minutes after assuming the upright position, and disappears or improves in less than 30 minutes after resuming the recumbent position⁷. Neck stiffness, tinnitus, hypoacusis, photophobia, nausea are the other common symptoms and suggest meningeal irritation. Schievink described criteria for the diagnosis of CSF leaks and intracranial hypotension that rely on both well-established radiographic and clinical findings⁸.

MRI of the head has proven to be an extremely useful investigation that confidently diagnoses SIH. Characteristic features of SIH include subdural fluid collection, enhancement of the pachymeninges, engorgement of venous structures, pituitary hyperemia and sagging of the brain⁹.

Computed tomography (CT) scan, although not as conclusive as MRI, can also be a useful investigation in emergency setting. It can demonstrate collection of fluid in the subdural space, herniation of cerebellar tonsils, collapse of cerebral ventricles and obliteration of the subarachnoid cisterns¹⁰.

It would be ideal to visualize the site of CSF leak but this is not possible in majority of cases even with an MRI of spine. It may, however, show signs of SIH which might include dilatation of veins in epidural and/or intradural space, enhancement of duramater, meningeal diverticula and CSF collection in the

extrathecal and retrospinal spaces. In some cases, MR myelogram, in contrast to conventional MRI may localize the CSF leak¹¹.

If spinal puncture is performed, the CSF opening pressure is typically less than 6 cms of H₂O (reference range, 6.5-19.5 cms of H₂O) but can be immeasurable or even negative. However, some patients have consistently normal CSF opening pressures³.

Most cases of SIH are self-limiting and respond well to bed rest, aggressive oral hydration, caffeine and glucocorticoids, or mineralocorticoids⁹.

The mainstay of treatment for SIH is the injection of autologous blood into the spinal epidural space, the so-called epidural blood patch¹². This is believed to seal the leak by tamponading the dura and is effective

in providing immediate symptomatic relief in about one third of patients⁹. In patients where the epidural blood patch provides temporary or incomplete relief, a second blood patch can be performed. In cases of complete failure, fibrin glue or surgical repair is indicated⁹.

Symptoms associated with SIH can be debilitating if timely action is not taken and can lead to brain herniation. Early diagnosis with appropriate management is an important aspect of SIH. Clinical features and radiographic findings can vary, with diagnosis largely depending on clinical suspicion, MRI and myelography. Multiple options for the treatment are available but with limited evidence for their effectiveness. Much is needed to be learnt about SIH to provide better care to patients with this disorder.

References

1. SCHALTENBRAND G: Neure anshaungun zur pathophysiologie der liquorzirkulation. *Zentrabl Neurochir*; 1938, 3:290-300.
2. Headache Classification Committee of the International Headache Society: Classification and diagnostic criteria for headache disorders, cranial neuralgias and facial pain. *Cephalalgia*; 1988, 8:1-96.
3. SCHIEVINK WI, MEYER FB, ATKINSON JLD, MOKRI B: Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension. *J Neurosurg*; 1996, 84:598-605.
4. SCHIEVINK WI, TORRES VE: Spinal meningeal diverticula in autosomal dominant polycystic kidney disease. *Lancet*; 1997, 349:1223-1224.
5. DAVENPORT RJ, CHATAWAY SJ, WARLOW CP: Spontaneous intracranial hypotension from a CSF leak in a patient with Marfan's syndrome. *J Neurol Neurosurg Psychiatry*; 1995, 59:516-519.
6. SCHIEVINK WI, MAYA MM, MOSER F, TOURJE J, TORBATI J, TORBATI S: Frequency of spontaneous intracranial hypotension in the emergency department. *J Headache Pain*; 2007, 8:325-8.
7. SCHIEVINK WI: Spontaneous spinal cerebrospinal fluid leaks: a review. *Neurosurg Focus*; 2000, 15:9(1):e8.
8. SCHIEVINK WI, MAYA MM, LOUY C, MOSER FG, TOURJE J: Diagnostic criteria for spontaneous spinal CSF leaks and intracranial hypotension. *AJNR Am J Neuroradiol*; 2008, 29:853-6.
9. SCHIEVINK WI: Spontaneous spinal cerebrospinal fluid leaks. *Cephalalgia*; 2008, 28:1347-1356.
10. SCHIEVINK WI, MAYA MM, TOURJE J, MOSER FG: Pseudo-subarachnoid hemorrhage: a CT-finding in spontaneous intracranial hypotension. *Neurology*; 2005, 65:135-7.
11. KATRAMADOS A, PATEL SC, MITSIAS PD: Non-invasive magnetic resonance myelography in spontaneous intracranial hypotension. *Cephalalgia*; 2006, 26:1160-4.
12. BERROIR S, LOISEL B, DUCROS A, BOUKOBZA M, TZOURIO C, VALADE D, BOUSSER MG: Early epidural blood patch in spontaneous intracranial hypotension. *Neurology*; 2004, 63:1950-1951.