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Spontaneous Intracranial Hypotension

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Abstract

Background: Spontaneous intracranial hypotension (SIH) is a rare condition characterized by a cerebrospinal fluid (CSF) leak, leading to low CSF pressure within the skull. The exact etiology is not always clear, but potential contributing factors include connective tissue disorders, trauma, spinal procedures, and structural deformities. Early diagnosis and management based on presenting signs, symptoms, and timely investigations can prevent progression to more serious complications. *Case Report*: We present the case of a 39-year-old male with no known co-morbidities who reported a three-week history of postural headache. The headache worsened upon standing and improved when lying down. Lumbar puncture revealed an opening CSF pressure of 40 mm Hg. Cranial and spinal MRI showed pachymeningeal thickening, subdural effusion, and cerebellar tonsillar herniation. Conservative management, including bed rest, led to symptom resolution within two weeks. *Conclusion*: This case emphasizes the importance of recognizing the clinical features, diagnostic methods, and management options for SIH. Imaging and lumbar puncture were crucial for diagnosis, and conservative treatment effectively alleviated symptoms. This case contributes to the medical understanding and support for individuals affected by SIH.

Keywords: Cerebrospinal Fluid, Diplopia, Headache, Lumbar Puncture, Subdural Effusion.

Introduction

Spontaneous intracranial hypotension (SIH) is caused by a cerebrospinal fluid (CSF) leak, typically at the spinal level, leading to reduced CSF volume around the brain and spinal cord. Causes of CSF leaks include congenital factors, dural tears, herniated discs, osteophyte projections, and surgeries. There is also an association with genetic disorders like Ehlers-Danlos syndrome and Marfan syndrome. SIH is more common in females, with peak incidence around 40 years of age, and an estimated incidence of 5 per 100,000 [1].

Patients usually present with orthostatic headaches, though non-orthostatic headaches can also occur [2]. Other symptoms include nausea, vomiting, diplopia, and vertigo. Complications may include cerebral venous sinus thrombosis and bibrachial amyotrophy. Diagnosis is guided by the International Classification of Headache Disorders (ICHD-3) **[Table 1]**, requiring evidence of CSF leakage on imaging or low CSF pressure on lumbar puncture [3]. Key diagnostic tools include Brain CT, Cranial MRI, and CT myelography (CTM) [4]. Recent advances in diagnosis involve transorbital ultrasound and serum biomarkers like serum transferrin [5,6].

This case report describes a 39-year-old patient with spontaneous intracranial hypotension (SIH) who presented with postural headache and responded well to conservative treatment.

Case Report

A 39-year-old male with no known co-morbidities presented to the neurology clinic with a 3-week history of headache. The headache worsened upon standing and walking, particularly in the

evening, and was localized to the occipital area, often radiating to the bifrontal region. The headache intensified with coughing, sneezing, and the Valsalva maneuver but improved upon lying down and reappeared when upright. A lumbar puncture revealed an opening CSF pressure of 40 mm Hg. The patient had no history of fever, neck stiffness, nausea, vomiting, limb weakness, visual disturbances, leg or back pain, head injury, or previous surgeries.

Cranial and spinal MRI showed pachymeningeal thickening in the bilateral frontoparietal, temporal, and occipital convexities with prominent subdural space **[Fig.1,2]**. There was bilateral frontoparietal subdural effusion, dilated superior sagittal sinus, and bilateral transverse sinuses. Cerebellar tonsillar herniation was noted about 3 mm below the basion line **[Fig.3]**, with an obliterated cisterna magna and reduced CSF space around the bilateral optic nerves **[Fig.4]**. The PNS examination revealed rightward DNS with inferior turbinate hypertrophy. There was no evidence of

Table 1: *Modified ICHD-III Diagnostic Criteria for Spontaneous Intracranial Hypotension**

A. Any headache attributed to low CSF pressure or CSF leakage that meets criterion C, below
B. Either or both of the following:
- Low CSF pressure (< 60 mm Hg)
- Evidence of CSF leakage on imaging
C. Headache that developed in temporal relation to the low CSF pressure or CSF leakage or that lead to its discovery
D. Headache not better accounted for by another ICHD-III diagnosis
*This criteria also applies to patients who do not have headache but whose symptoms are best elucidated by spontaneous intracranial hypotension. Headache pointing towards SIH cannot be diagnosed in a patient who within the prior month, has had trauma or a procedure known to produce CSF leak.

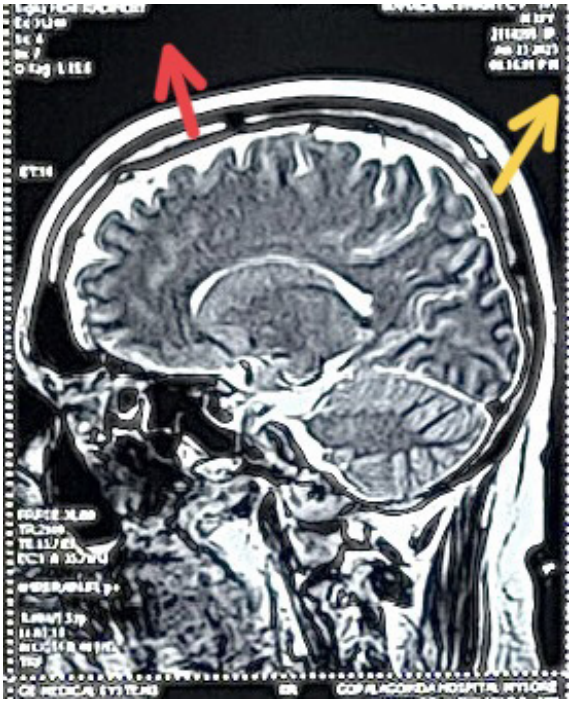

Fig.1: *Pachymeningeal enhancement (red arrows) and prominent subdural space (yellow arrows).*

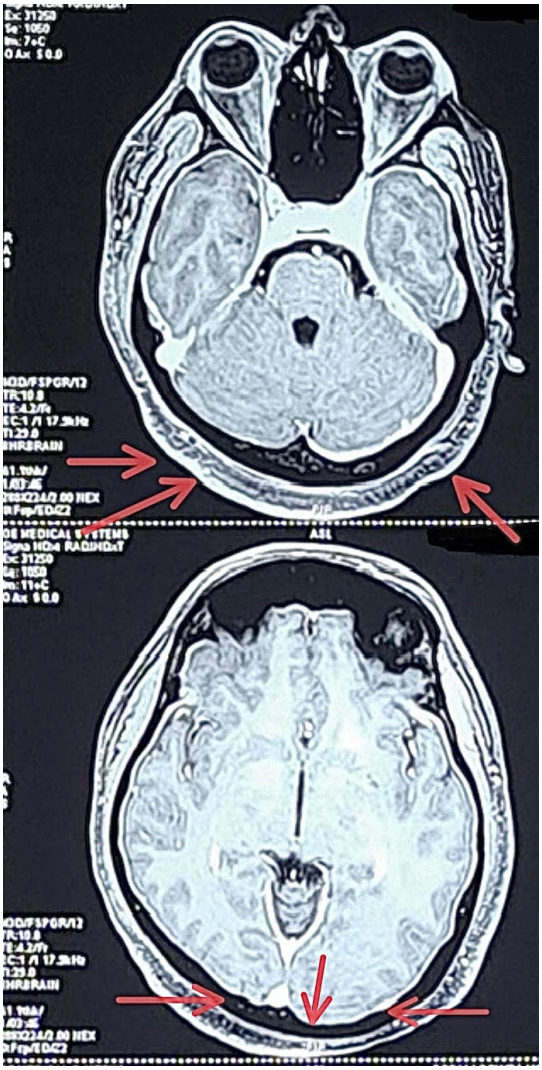

Fig.2: *Pachymeningeal enhancement (red arrows) and prominent subdural space (yellow arrows).*

intracranial bleeding, masses, ventriculomegaly, or pituitary abnormalities. The spinal MRI was normal **[Fig.5]**, and cisternography was deemed unnecessary.

The patient was advised to maintain bed rest for 2 weeks, after which his symptoms significantly improved. There were no adverse or unanticipated events reported during the treatment.

Discussion

Spontaneous intracranial hypotension (SIH) can result from minor trauma or weaknesses in the dural sac, often due to spontaneous dural dehiscence or degenerative spinal diseases. SIH can present with symptoms that mimic other conditions, making differential diagnosis crucial. Symptoms such as nausea, vomiting, photophobia, and posterior neck pain/stiffness can resemble subarachnoid hemorrhage or infectious meningitis. Additionally, high protein content and pleocytosis in CSF analysis can mislead physicians towards a meningitis diagnosis, while cerebellar tonsillar herniation might be confused with Chiari I malformation [7].

Patients with SIH typically present with headaches that change with posture, worsening when upright and alleviating when lying down. Intracranial hypotension can be classified as either spontaneous or secondary, with the latter often resulting from iatrogenic causes like lumbar puncture, leading to a CSF leak.

Many SIH patients experience mild symptoms that may resolve without intervention. Conservative management, including bed rest, is often effective, as was the case with our patient, whose symptoms resolved within two weeks. Lying supine reduces CSF pressure at the leak site, facilitating healing. Treatments such as caffeine and theophylline can reduce symptoms by inducing arterial contraction and decreasing intracranial blood flow and venous engorgement. If conservative measures fail, an epidural blood patch can be used to seal the CSF leak, with the

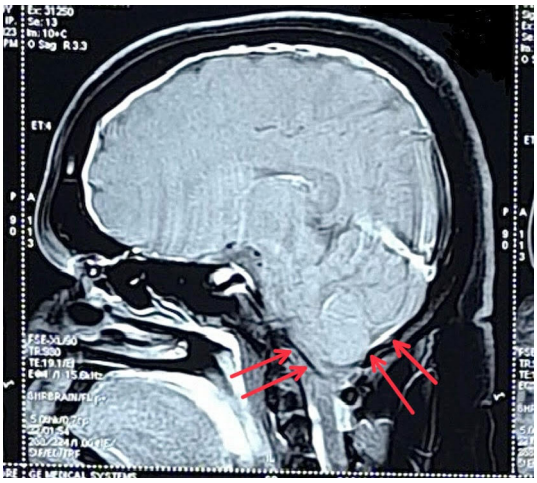

Fig.3: *Evidence of cerebellar tonsillar herniation.*

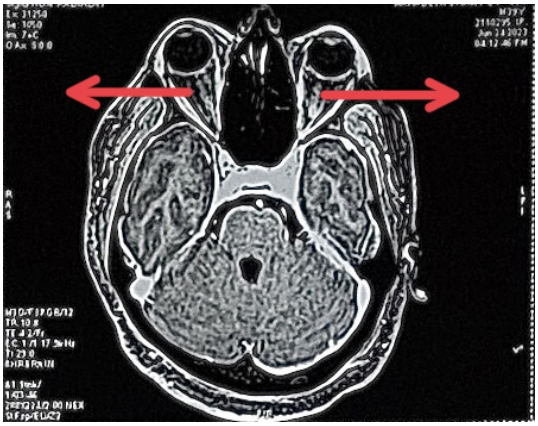

Fig.4: *Effacement of perioptic CSF space.*

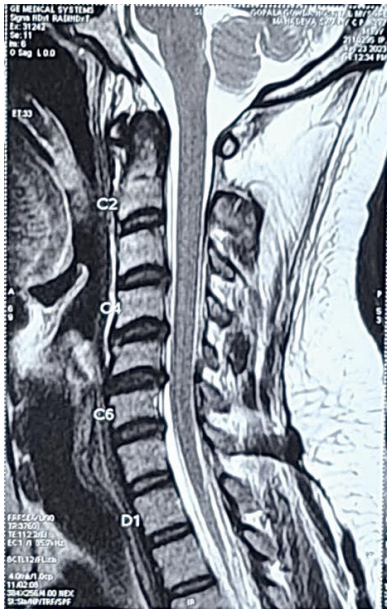

Fig.5: *Normal spinal MRI.*

injected blood forming a clot that helps close the leak [6]. Should these less invasive methods prove ineffective, surgical options may be considered, including the ligation of leaking meningeal diverticula, direct repair of dural tears, packing the epidural space with fibrin glue, or strengthening the dura via duroplasty [2,5].

Conclusion

This case highlights the clinical features, diagnostic methods, and management of SIH. Imaging and lumbar puncture were critical in diagnosing SIH, and conservative management effectively relieved the patient's symptoms without further intervention. Recognizing SIH's clinical presentation and conducting thorough MRI assessments are essential for accurate diagnosis, preventing unnecessary invasive procedures, and enhancing medical understanding and support for affected individuals.

Contributors: SP was directly involved in the patient care of this patient, he obtained consent, collected the data used in the case report. ND, AB: drafting, discussion, literature search and critical revision of the manuscript. They also acted as the study guarantors. All authors approved of the final version of this manuscript and are responsible for all aspects of this study.
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