

## RADIOLOGICAL FEATURES OF SPONTANEOUS INTRACRANIAL HYPOTENSION (SIH) AND ITS MANAGEMENT WITH LUMBAR EPIDURAL AUTOLOGOUS BLOOD PATCH: A CASE REPORT

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### ABSTRACT

Spontaneous intracranial hypotension (SIH) is an important but one of the rarest causes of headache in young and middle- aged people. Its incidence is about five per 100,000 people [1], with peak at 40 years of age and twice more common in women than men. The classic symptom of SIH patient is postural or orthostatic headache which aggravates on standing position and is relieved on lying down position.

We reported a rare case of spontaneous intracranial hypotension (SIH) in fourty year old gentleman who presented with classic symptoms of orthostatic hypotension. The patient was evaluated with computed tomography (CT), non-contrast & contrast enhanced magnetic resonance imaging (MRI). CT myelography confirmed the CSF leaks at multiple spinal nerve root levels.

The patient was treated with therapeutic lumbar epidural blood patch (LEBP) by injection of 10-20mL autologous blood into the epidural space when all available conservative treatments were failed. There was significant improvement of orthostatic headache of patients with in few days of LEBP.

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### INTRODUCTION

Spontaneous intracranial hypotension (SIH) is one of the rarest causes of headache in young and middle- aged people with incidence of about five per 100,000 people [1]. It peaks at 40 years of age and twice more common in women than men.

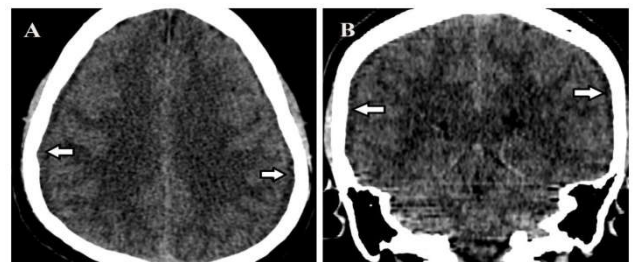
The classic symptom of SIH patient is postural or orthostatic headache which aggravates on standing position and is relieved on lying down position. The syndrome of spontaneous intracranial hypotension (SIH) is due to low cerebrospinal fluid (CSF) volume resulting from leakage of CSF from the thecal sac to extrathecal sac space within or along the spinal canal [2, 3].

In this article, we have reported radiological features of spontaneous intracranial hypotension (SIH) in fourty year old gentleman who presented with classic symptoms of orthostatic hypotension. On CT and MR imagings, there were classic findings of subdural hygroma and diffuse pachymeningeal enhancement. CT myelography confirmed the CSF leaks at multiple spinal nerve root levels. The patient was treated successfully with therapeutic epidural blood patch by injection of autologous blood into the epidural space.

### CASE REPORT

A forty year old gentleman presented to the department of neurology with chief complaint of orthostatic headache for few months. The headache was worsening on standing position and improved in a recumbent position. No past history of convulsion, head trauma or medical conditions noted. Patient took treatment for chronic headache, but without any relief.

General and systemic examination of patients was unremarkable. Otolaryngological and ophthalmologic examinations were normal. Neurological examination revealed normal cranial nerves, motor and sensory function. Serum biochemistry values were within normal limits.



**Figure 1** Fourty year old male patient with spontaneous intracranial hypotension. Non-contrast computed tomography Axial (A) and Coronal (B) image show hypodense subdural hygroma in bilateral cerebral convexities (arrow).

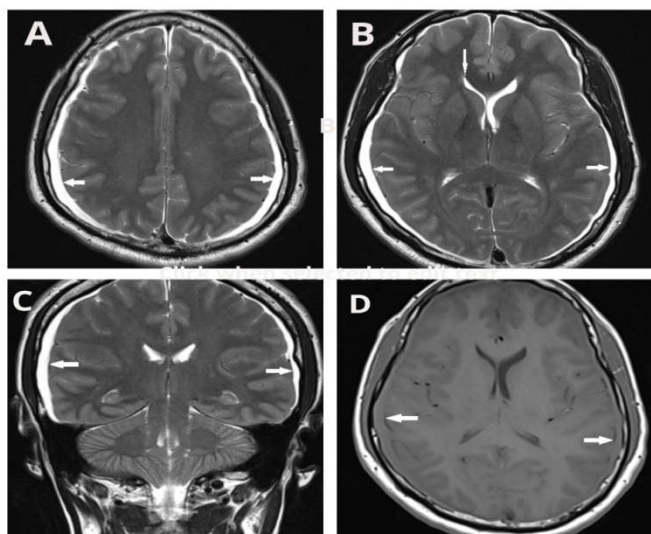
Non contrast computed tomography (NCCT) of brain revealed fluid density collections suggesting subdural hygroma in

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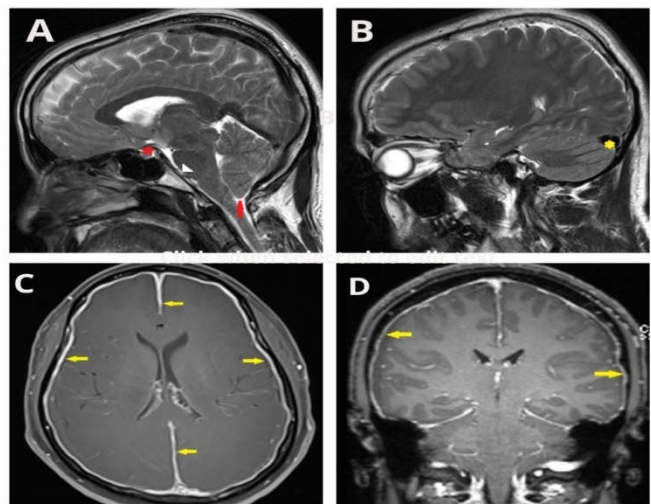
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bilateral cerebral convexities with effaced cerebralsulci (Figure 1A, 1B). Brain parenchyma, orbits and paranasal sinuses are grossly normal. Non contrast and contrast-enhanced magnetic resonance imaging (MRI) of brain was advised for further evaluation.

On MRI, bilateral symmetrical cerebral convexity hyperintense signal collection on T2-weighted images (Figure 2A, 2B, 2C). Isointense signal on T1-weighted images noted (Figure 2D). There was tonsillar herniation (>5mm), effaced prepontine cisterns, reduced ponto-mesencephalic angle and indentation over corpus callosum splenium due to sagging of brain. Above findings represent intracranial hypotension (Figure 3A) best appreciated on T2 sagittal images. There was venous engorgement of the superior sagittal and transverse sinuses with rounding of transverse sinus (Figure 3B).

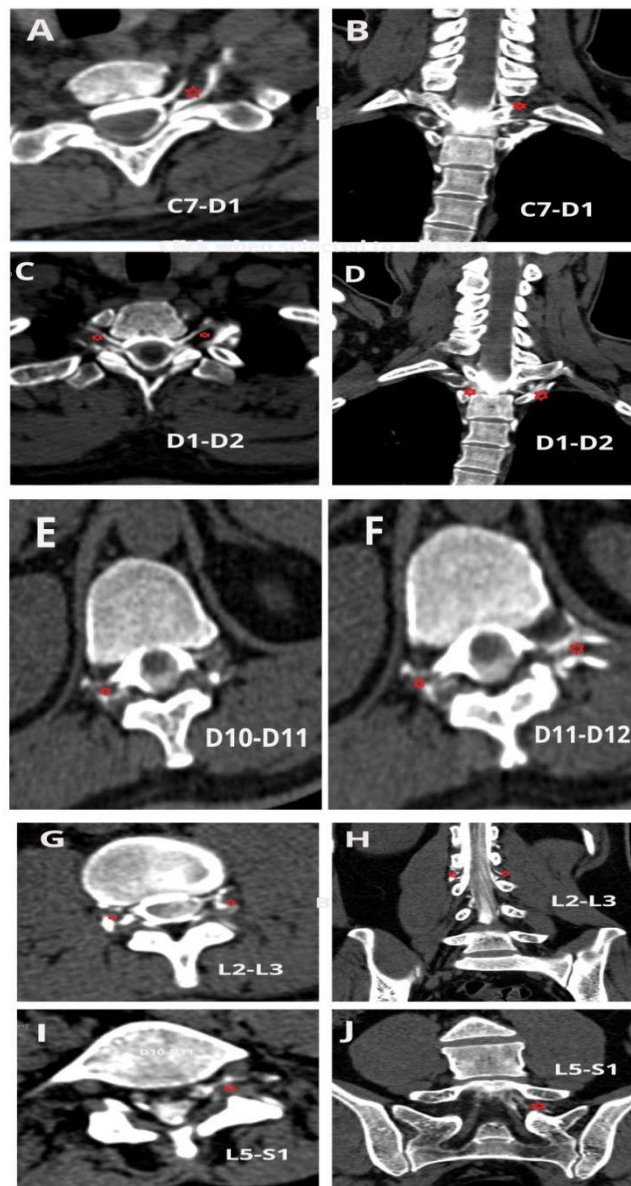


**Figure 2** Forty year old male patient with spontaneous intracranial hypotension. Non-contrast magnetic resonance T2 weighted Axial (A, B) and Coronal (C) image show hyperintense subdural hygroma in bilateral cerebral convexities (arrow) with effacement of sulci. T1 weighted Axial (D) image show isointense subdural hygroma in bilateral cerebral convexities (arrow).



**Figure 3** Forty year old male patient with spontaneous intracranial hypotension. Non-contrast magnetic resonance T2 weighted Sagittal (A) image show low-lying cerebellar tonsils (red arrow), downward retraction of pituitary infundibulum (red star), effacement of prepontine cisterns and flattening and straightening of the ventral pons (triangle). Coronal image (B) reveals venous engorgement with rounding of transverse sinus (yellow star). Contrast enhanced magnetic resonance T1 weighted Axial (C) and Coronal (D) image show diffuse smooth thickening and uniform intense enhancement of pachymeninges.

On post contrast T1-weighted images, diffuse smooth thickening and uniform intense enhancement of dura suggesting pachymeningeal enhancement was noted. (Figure 3C, 3D). Bilateral optic nerve sheath complexes were normal. No obvious site of CSF leak found.



**Figure 4 (A-J):** Forty year old male patient with spontaneous intracranial hypotension. Computed tomography myelogram images of Cervico-Dorsal-Lumbar spine images show extra-dural fluid collections consistent with CSF leaks at multiple levels (red stars). Axial (A) and Coronal (B) at C7-D1 level. Axial (C) and Coronal (D) at D1-D2 level. Axial at D10-D11 (E) & D11-D12 (F) level. Axial (G) and Coronal (H) at L2-L3 level. Axial (I) and Coronal (J) at L5-S1 level.

Diagnostic CT myelogram was advised to evaluate the cause of hypotension. After infusion of 50% diluted non-ionic iodinated contrast media (350mmol/mg) into epidural sac by lumbar puncture, scanning from base of skull to coccyx was done in left lateral, right lateral, supine and prone positions.

CT myelogram revealed extra-theal contrast media collection at multiple nerve roots levels along cervical, dorsal and lumbar spine. These sites were representing cerebrospinal fluid (CSF) leakage (Figure 4A-4J).

The diagnosis of SIH was made on the basis of typical headache symptoms, characteristic MRI and CT myelogram findings. The patient was put on conservative treatments, but

condition remained unchanged. Hence trial of therapeutic epidural blood patch was done with injection of 10-20mL autologous blood into the epidural space. In next few days, there was significant improvement of orthostatic headache and he was put on 3-month follow up.

## **DISCUSSION**

Spontaneous intracranial hypotension (SIH) is an important but one of the rarest causes of headache in young and middle-aged people. Its incidence is about five per 100,000 people [1], with peak at 40 years of age and twice more common in women than men.

The cause of intracranial hypotension is either spontaneous (primary) or related to injury (secondary of cranio-spinal axis). The syndrome of spontaneous intracranial hypotension (SIH) is due to low cerebrospinal fluid (CSF) volume resulting from leakage of CSF from the thecal sac to extrathecal space within or along the spinal canal [2, 3]. Most of the times, the exact cause of spontaneous CSF leak is not identifiable, however occult CSF leak through small defects in the meninges is presumed to be pathogenesis of decrease in CSF volume and pressure. The opening pressure on lumbar puncture usually is low (0-70mm H<sub>2</sub>O).

Patient with intracranial hypotension complains of various non-specific symptoms such as nausea, vomiting, neck pain, visual and hearing disturbances, and vertigo. However, the classic symptom of SIH patient is postural or orthostatic headache which aggravates on standing position and is relieved on lying down position. The low CSF pressure results in downward displacement of the brain and stretching of pain-sensitive dura due compensatory dilation of venous structures. These various non-specific symptoms may lead to misdiagnosis and delay the effective treatment. Hence proper history and high suspicion is crucial for diagnosis of SIH.

The diagnosis of spontaneous intracranial hypotension due to spontaneous spinal CSF leak according to international classification of headache disorders requires the following: the orthostatic headache with at least one additional symptoms (tinnitus, neck stiffness, hypoacusia, photophobia, or nausea) and low CSF pressure evidence on contrast enhanced MRI, x-ray myelography, CT myelography, cisternography or CSF opening pressure [4].

The assessment of the underlying spinal pathologic condition of SIH is done by dynamic CT myelography, X-ray myelography, conventional MR imaging and MR myelography. Contrast enhanced brain MRI and CT myelography are choice of imaging in early evaluation of SIH. But dynamic CT myelography is considered to be gold standard for diagnosis of spinal CSF leak. The imaging findings may be normal in some SIH patients.

Subdural fluid collection or subdural hygroma in bilateral cerebral and cerebellar convexities is most common MR imaging finding in 50% of cases [5]. Diffuse, smooth and intense pachymeningeal (dural) enhancement on gadolinium enhanced MRI involving both supratentorial and infratentorial compartments bilaterally [6] are the most specific abnormality. Enlargement and rounded appearance of flow-void of dural venous sinuses on MRI is one of characteristic imaging finding [9] appreciated on sagittal and coronal images. On sagittal MRI, venous distention sign involves convex bulging of the inferior border of the dominant transverse sinus [9]. Brainstem

slumping or sagging best appreciated on sagittal MRI sign is specific for intracranial hypotension [6] and noted in about 51% of patients. The imaging signs of brainstem slumping described in the literatures are effacement of ventricular system, supra-sellar and prepontine cisterns; flattening and straightening of the ventral pons, compression of the optic chiasm over the sella and caudal herniation of the cerebellar tonsils into spinal canal [6, 9]. In our cases, almost CT and MR imaging findings are correlated with available literatures.

CT myelography (CTM) provides very good spatial resolution and a good imaging modality for identifying CSF leak site after injection of non-ionic contrast material into the subarachnoid space. Alternatively, MR myelography (MRM) can be used to localize spinal CSF leaks. However, CTM is good to localize the slow CSF leaks [7] and dynamic CTM is best for fast CSF leak localization [8]. CT myelographic appearance of CSF leaks from nerve root sleeve diverticulum depends on the size of the dural defect and the volume of leak. Most of the times, slow leak is quite subtle and may be missed. It generally results collection of small volume of contrast within the neural foramen adjacent to nerve root sleeve. High flow CSF leaks results in collection of large volume of contrast in extrathecal region adjacent and along nerve root sleeve [8, 10]. In our case, extrathecal CSF due to leaks are noted at multiple spinal levels.

Initially, the treatment of SIH is conservative with aim to maintain the CSF pressure. Bed rest, adequate hydration, caffeine and steroids are conservative measures. Lumbar epidural blood patch (LEBP) is one of promising approach for the treatment when conservative treatment fails [11]. Non-targeted LEBP has variable success in cases of SIH and many of patients experience symptom relief of intracranial hypotension within minutes [11]. Long term relief of symptoms presumably occurs due to sealing of dural defect by the LEBP and gradual restoration of CSF volume. Surgery is effective for few of patients with large dural defect refractory conservative measures, LEBP, and epidural fibrin glue patching [12]. In this case, lumbar epidural blood patch was tried with significant symptomatic relief of orthostatic hypotension.

## **SUMMARY AND CONCLUSION**

Spontaneous intracranial hypotension (SIH) is one of rarest cause of orthostatic hypotension due to spinal CSF leak, diagnosed with typical clinical symptoms, imaging findings and low CSF opening pressure. Bilateral subdural hygroma with diffuse pachymeningeal enhancement, tonsillar herniation and engorged dural sinuses are typical imaging findings on CT and contrast enhanced MRI. Dynamic CT myelography is gold standard to localize the site and volume of spinal CSF leak. Epidural blood patch has promising symptomatic relief in most of patients with SIH. Only in a few of refractory cases, surgery is needed to close the dural defect.

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