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Case Study of Spontaneous Intracranial Hypotension

Lipika Gupta[1], Archit Gupta[2], Deepak Varshney[3]

[1]Post Graduate Trainee, Department of Radiodiagnosis, NEIGRIHMS, Shillong, India
Email id: *guptalipika94[at]gmail.com*

[2]Assistant Professor, Department of Radiodiagnosis, SNMC, Agra, Uttar Pradesh, India
Corresponding author Email id: *architgrmc[at]gmail.com*
Phone: 8828275864, 9926295237

[3]Associate Professor, Department of Radiodiagnosis, SAMC and PGI, Indore, India
Email id: *deelovi[at]yahoo.coin*

Abstract: ***Spontaneous intracranial hypotension is a condition that clinically presents with an orthostatic headache caused by low cerebrospinal fluid pressure (CSF). The other symptoms are mainly due to traction on the cranial and spinal nerves secondary to descent of the brain caused by low CSF pressure. It hascharacteristic magnetic resonance imaging (MRI) findings among which effacement of the suprasellar cistern, engorgement of the venous sinuses and pachymeningeal enhancement have the highest diagnostic sensitivity. This condition is in almost all cases is caused by spinal cerebrospinal fluid leaks.***

Keywords: intracranial hypotension; magnetic resonance imaging; pachymeningeal enhancement; dural sinus engorgement

1. Introduction

Intracranial hypotension (IH), first described in 1938 by Schaltenbrand [1], is an important condition that has characteristic clinical and MRI findings. It is an uncommon condition, with an estimated incidence of 5 per 100, 000 per year [2], and it usually affects young to middle - aged adults, with a female predominance [3].

Intracranial hypotension is of either primary (spontaneous intracranial hypotension – SIH) or of secondary origin e. g. iatrogenic or traumatic. Spontaneous intracranial hypotension is believed to occur as a result of trivial trauma, weakness in the dural sac due to spontaneous dehiscence of the dura and dural tears caused by degenerative causes. An association has also been found with the connective tissue disorders such as Marfan and Ehlers - Danlos syndromes [2, 4, 5]. Secondary intracranial hypotension may be caused by injury of the duramater, e. g. following lumbar puncture, spinal anaesthesia, cranial or spinal surgery, placement of ventriculo - peritoneal (VP) shunts, and as a result of craniospinal trauma [2, 4].

CSF hypovolemia plays a major role in the development of SIH as compared to CSF hypotension as many patients may have normal CSF pressure despite having typical symptoms of SIH. [6, 7] Approximately 10% of the estimated total CSF volume has to be reduced to induce orthostatic headache. [8] Headache occurs due to stretching of pain - sensitive intracranial structures due to descent of brain which is more in the standing position & secondary to venous engorgement to compensate for the lost CSF as per Monro–Kellie doctrine. [9]

Magnetic resonance imaging is very useful in the diagnosis because of its characteristic findings [10]. Certain measurements are also done for diagnosis of IH which include Pontomesencephalic angle, Mamillopontine distance and Lateral ventricular angle. In a study by **Shah et al.,** Pontomesencephalic angle is defined as angle between line drawn along anterior margin of midbrain and anterior superior margin of pons. Mean value in patients with intracranial hypotension was 41.2° (SD, ± 17.4°). Mamillopontine distance is defined as distance between inferior aspect of mamillary bodies to superior aspect of pons. Mean value in patients with intracranial hypotension was 4.4 mm (SD, ± 1.8). Lateral ventricular angle is defined as angle between medial margins of right and left lateral ventricles. This angle was measured on coronal imaging at level of fornices. Mean value in patients with intracranial hypotension was 130.1° (SD, ± 9.8°). [11]

SIH needs to be differentiated form various radiological mimickers like Chiari type 1 malformation, subdural fluid collection and diseases associated with dural thickening like neurosarcoidosis, tuberculosis, autoimmune disorders. The various important complications also need to be identified on imaging i. e subdural hematomas, cerebral venous thrombosis and superficial siderosis.

2. Case Report

A 48 year old man presented with symptoms of headache and neck pain for the past 4 days with imbalance while bending forward. There was no history of trauma, fever or surgical intervention. There was no relief with medications. Neurological examination was normal. MRI was done on a 1.5 - T scanner (AVANTO, Siemens AG, Germany) which showed bilateral fronto - parietal subdural hygroma appearing hypointense on T1 - weighted and hyperintense on T2 - weighted images (Figure 1). Sagittal and axial images showed sagging/slumping of midbrain with effacement of perimesencephalic cisterns, right uncal herniation and mild inferiorly displaced cerebellar tonsil [Figure 1 (c) (d)]. The

right transverse and sigmoid sinuses were engorged [Figure 2 (a)] with pachymeningeal enhancement seen on post contrast study [Figure 2 (b)]. Various diagnostic intracranial angles were calculated which were also in favour of diagnosis of intracranial hypotension (Figure 3). These findings along with the clinical history were suggestive of spontaneous intracranial hypotension. MRI of the spine did not show any abnormality. The patient was put on conservative treatment.

3. Conclusion

Intracranial hypotension (IH) is a benign, uncommon, and usually a self - limiting condition caused by low cerebrospinal fluid (CSF) pressure, usually due to CSF leakage. The dominant clinical feature is an orthostatic headache. Other common clinical features include fever, nausea, vomiting, and tinnitus. Magnetic resonance imaging (MRI) plays an important role in the diagnosis and follow - up of such patients. Most prominent features of spontaneous intracranial hypotension on MR imaging include “brain sag” and diffuse pachymeningeal enhancement [12]. However, features like sagging of the brain, pituitary enlargement, pachymeningeal enhancement and subdural fluid collections are considered as specific MRI findings [13]. Intracranial hypotension can mimic other conditions, hence, accurate diagnosis is important, because misdiagnosis may lead to unnecessary procedures and prolonged morbidity.

Conflicts of interest: The authors declare that they have no conflicts of interest

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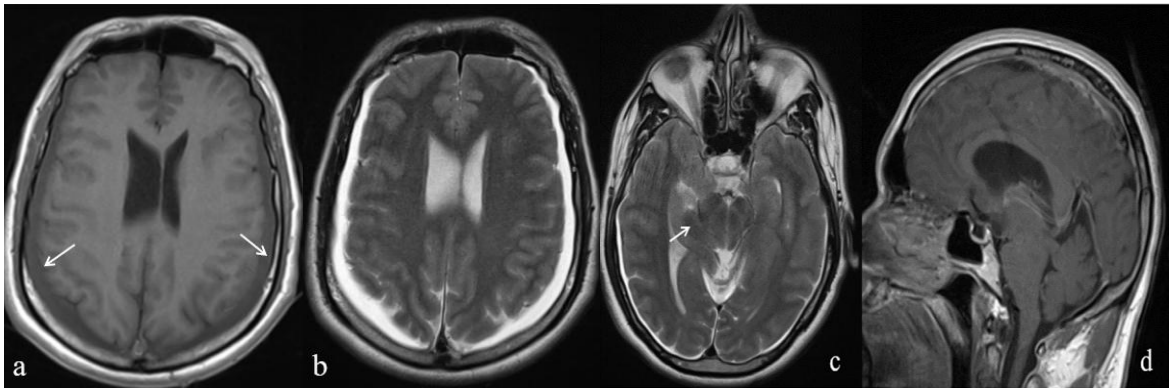

Figure 1: Axial (a) T1 - weighted (b) T2 - weighted images of the brain showing bilateral fronto - parietal subdural hygromas appearing hypointense on T1 - weighted (white arrows) and hyperintense on T2 - weighted images, (c) Axial T2 - weighted image showing effacement of perimesencephalic cisterns, right descending transtentorial uncal herniation (white arrow) and (d) Sagittal T1 - weighted post contrast image of the brain showing sagging of midbrain and inferiorly displaced cerebellar tonsil.

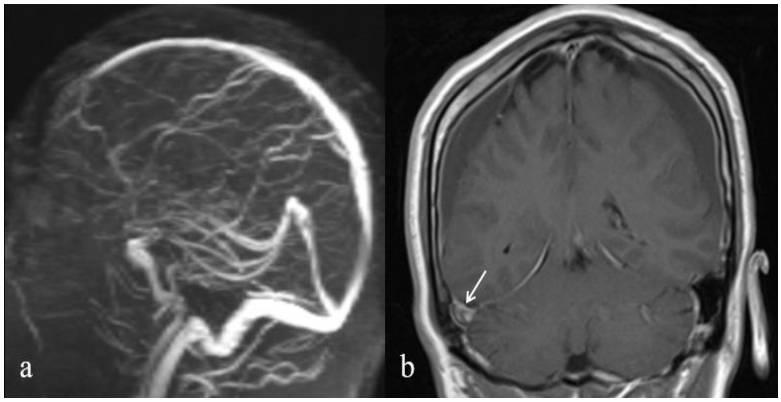

Figure 2: (a) Sagittal MRV image showing engorged right sided dural veinous sinus and (b) Coronal T1 - weighted post contrast image of the brain showing smooth pachymeningeal enhancement and engorged right dural veinous sinus (white arrow).

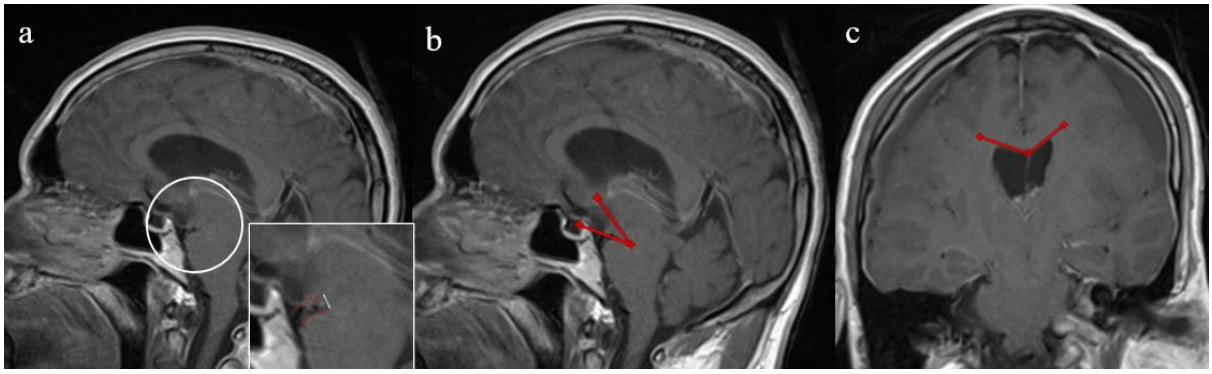

Figure 3: Diagnostic intracranial angles in patient with intracranial hypotension. (a) Mamillopontine distance is narrowed to 2.3 mm as measured on PACS using standard ruler tool. (b) Pontomesencephalic angle is narrowed to 33° as measured on PACS using standard angle - measuring tool. Note also low - lying cerebellar tonsils and sagging of brainstem (c) Lateral ventricles ventricular angle is narrowed to 124.2°.