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Pictorial Review

Intracranial hypotension: An uncommon entity with common presentation

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ABSTRACT

Headache disorders are among the most common presenting complaints in any neurology outpatient department. International Headache Society classifies headaches as “primary” or “secondary”. The causes of secondary headaches are varied and intracranial hypotension is one of them. It typically presents clinically with postural headaches but most of the times, its diagnosis is delayed, as it is an uncommon cause and there is poor awareness among the medical fraternity about this condition. Imaging, especially magnetic resonance imaging (MRI), plays a crucial role in the diagnosis of intracranial hypotension by not only confirming the diagnosis but also detecting the cause in some cases. This case series tries to highlight the MRI findings of intracranial hypotension in three cases with different etiologies.

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Introduction

Headache is one of the most common presenting complaints in any neurology outpatient department. Headache disorders are associated at times with disability, poor quality of life, and financial burden for the patient. International Headache Society classifies headaches as “primary” or “secondary”.¹ Primary headaches do not have any underlying cause or pathology and are benign and recurrent in nature. Secondary headaches have an underlying cause which may be an infection, vascular disorders, cerebral bleed, tumors, or head injury. History taking and thorough neurological examination are generally enough to arrive at a conclusion, whether the

headache is primary or secondary. Neuroimaging is utilized to evaluate the suspected cases of secondary headaches. We present here three cases of headaches which were diagnosed on magnetic resonance imaging (MRI) as intracranial hypotension (IH).

Case reports

Case 1

A 19-year-old male patient presented with history of severe headache on and off for the last 4 months which was localized to occipital region and nape of neck. The patient was apparently asymptomatic till 4 months back when he sustained a flexion injury to neck while playing volleyball and developed severe headache 4 h later which was relieved

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Fig. 1 – (a) Axial T2WI showing anteroposterior elongation of mid brain (black arrow) and rounded superior sagittal sinus (arrow). (b) Axial T2WI shows normal appearance of superior sagittal sinus having triangular configuration for comparison (arrow). (c) Sag T1WI image showing venous distension of dominant right transverse sinus as convex bulging of superior border (arrow).

by rest and hydration. The MRI brain and computerized tomography (CT) neck done a week later were reported as normal. For the next 4 months, the patient suffered multiple episodes of such severe headaches which were typically brought on when the patient was standing or sitting and was relieved by lying down. There was no association with physical exertion or time of the day. General examination was normal, and there was no neck stiffness, rigidity, or swelling. Routine laboratory investigations such as complete hemogram, urine routine examination (RE), blood sugar, urea, and serum creatinine were within normal limits. Central nervous examination revealed normal higher mental functions, with no

motor or sensory deficit, cranial nerve assessment was normal, and deep tendon jerks were normal. Fundus examination was normal. Based on the history and examination, clinical suspicion of IH was raised and the patient was subjected to MRI evaluation. MRI brain revealed anteroposterior elongation of mid brain and rounding of the superior sagittal sinus on axial T2W image and convex superior border of dominant right transverse sinus on Sag T1WI (Fig. 1a-c), and sagittal T1W image showed brainstem sagging, tonsillar herniation, flattening of central pons, and effacement of prepontine cistern (Fig. 2). The mamillopontine distance was 4 mm (Fig. 3) and the pontomesencephalic angle was 33.1°

Fig. 2 – Sag T1WI image showing tonsillar herniation (arrow), flattening of central pons, and effacement of prepontine cistern (broad arrow).

Fig. 3 – Sag T2WI showing mamillopontine distance of 4 mm.

(Fig. 4). Post-contrast axial and coronal images showed supra- and infratentorial pachymeningeal enhancement (Fig. 5a and b). These MRI brain findings were consistent with IH. In view of history of injury to neck, screening of cervical spine with Sag short tau inversion recovery (STIR) sequence was performed which showed extradural fluid intensity at the level of C2 (Fig. 6a). Axial Constructive Interference in Steady State (CISS) sequence at the level of C2 showed perithecral Cerebrospinal fluid (CSF) intensity suggestive of dural leak (Fig. 6b); however, dural defect could not be identified. CT cisternography was done which revealed nonuniform opacification of the thecal sac and failed to demonstrate the dural leak or extradural

collection (Fig. 6c). The case was diagnosed as post-traumatic IH.

Case 2

A 28-year-old female patient, a case of pineal parenchymal tumor (optd) with ventriculoperitoneal (VP) shunt in situ, presented with history of intermittent postural headaches 6 months after the operation. Headache was moderate in severity and not localized to any particular region, and it was aggravated on prolonged standing and relieved on lying down. There was no associated aura. General examination and

Fig. 4 – Sag T2WI showing the pontomesencephalic angle of 33.1°.

Fig. 5 – (a and b) Post-contrast axial T1W (FS) and coronal images showing supra- and infratentorial pachymeningeal enhancement.

Fig. 6 – (a) Screening Sag STIR sequence showing extradural fluid intensity at the level of C2 posteriorly (arrow). (b) Axial CISS sequence at the level of C2 showing perithecal CSF intensity suggestive of dural leak (arrow). (c) CT myelography at the level of C2 shows nonuniform opacification of thecal sac with linear opacification of the thecal sac by contrast anterolaterally (arrow).

routine laboratory investigations were normal. Central nervous system examination did not reveal any neurological deficit. All cranial nerves were normal. MRI was done as part of post-op follow-up for pineal parenchymal tumor. MRI revealed VP shunt in situ, and there was anteroposterior elongation of mid brain, rounding of the superior sagittal sinus and left subdural effusion seen on axial T2W images (Fig. 7a and b). There was sagging of brain seen as descent of iter (9 mm) (Fig. 8). The mamillopontine distance was 4.2 mm and the pontomesencephalic angle was 40°. However, there was no tonsillar herniation. Post-contrast axial images showed thick pachymeningeal enhancement with no evidence of residual or recurrent tumor in the pineal region (Fig. 9a and b). Screening of the spine did not reveal any abnormality. These MR findings were suggestive of IH probably due to shunt over function.

Case 3

A 41-year-old male patient presented with history of intermittent headaches since last 7 months which were mild to moderate in severity and localized to occipital region. The headache was aggravated on prolonged standing and typically relieved on lying down. There was no history of trauma or loss of consciousness. There was no aura associated with headache. General and CNS examinations were normal. Routine laboratory investigations were normal. CT scan head done 3 months earlier was reported as normal. In view of persistent symptoms, the patient was subjected to MRI brain for further evaluation. Axial T2W images showed anteroposterior elongation of mid brain, effacement of suprasellar cistern, rounding of the superior sagittal sinus, and bilateral subdural effusions (Fig. 10a and b). Sagittal T2W image revealed sagging

Fig. 7 – (a and b) Axial T2W images showing distended superior sagittal sinus (arrow) and left subdural effusion (broad arrow).

Fig. 8 – Sag post-contrast image shows low position of iter measuring 9 mm (normal 1.8–2 mm).

of brain stem with tonsillar herniation and reduced mamillo-pontine distance (Fig. 11). Axial contrast-enhanced image showed pachymeningeal enhancement (Fig. 12). The screening of spine did not reveal any fluid intensity in the extradural space on STIR images. The MRI findings were consistent with spontaneous intracranial headache.

Discussion

The main presenting feature of IH is the postural or orthostatic headache which usually occurs when the patient is sitting or

standing and the headache is relieved on lying down.² These headaches are generally localized to the occipital region but they may vary from patient to patient. The headache may vary in severity, but the pain is aggravated by Valsalva maneuver, jugular vein compression, and coughing, and is not relieved with analgesic agents. Anorexia, nausea, vomiting, and neck pain may also be sometimes associated with the headache which also get relieved with change in posture. Sometimes patients with IH may present with atypical symptoms such as diplopia, hearing changes, tinnitus, giddiness, visual blurring, and facial numbness. Rare presentations include coma, Parkinsonism, dementia, and quadriplegia.^{3,4}

Fig. 9 – (a and b) Axial post-contrast images at the level of lateral ventricles and pineal region show thick pachymeningeal enhancement (arrow) and no evidence of residual tumor.

Fig. 10 – (a and b) Axial T2WI showing anteroposterior elongation of mid brain (arrow), effacement of suprasellar cistern, rounding of the superior sagittal sinus and bilateral subdural effusions.

The surgical causes of IH include spinal surgery, VP shunting, craniospinal trauma, and CSF leak from lumbar puncture.⁵ Medical causes of IH include dehydration, uremia, diabetic coma, and hyperpnea.⁶ When it occurs in some patients without any identifiable cause, it is termed as spontaneous intracranial hypotension (SIH). Spontaneous dural tear which results in persistent CSF leak somewhere along the neuraxis is generally considered to be the cause of SIH.⁷ It typically occurs in middle ages (30–50 years) and has a predilection for females (M:F – 1:2). In this case series, first case was due to leakage of CSF following trauma to the neck, second

case was due to excessive shunting by the VP shunt, and the third case was due to SIH.

Patients with IH have decreased CSF opening pressure which is often less than 60 mm of H₂O at the time of lumbar puncture.⁸ But more than the low CSF pressure, it is the decrease in CSF volume which is responsible for development of IH as it has been found that some patients have normal CSF pressure but suffer from typical symptoms of IH.² The volume of CSF lost is compensated by increase in intracranial blood volume as per the Monro-Kelly hypothesis, which states that there is equilibrium between the volumes of brain tissue, CSF,

Fig. 11 – Sag T1W image showing tonsillar herniation (arrow) and reduced mamillopontine distance (4.1 mm).

and blood of the intracranial compartment. A reduction in volume of CSF results in compensatory dilatation of the venous sinuses.⁹ CSF hypovolemia and compensatory mechanisms lead to cascade of events which result in clinical and radiological manifestations of IH.¹⁰

The diagnosis of IH is suspected clinically when typical history of orthostatic headache is elicited but the imaging plays a vital role not only in confirming the diagnosis but also

Fig. 12 – Axial post-contrast image shows pachymeningeal enhancement.

on follow-up of these cases after conservative or definitive management. CT head is the least sensitive and does not help in clinching the diagnosis; however, it may demonstrate obliteration of basal cisterns, increased density in the basal cisterns termed as pseudo subarachnoid hemorrhage,¹¹ or in some cases dural thickening on post-contrast images.

MRI has transformed the way we diagnose IH. Majority of MRI findings are secondary to the compensatory mechanisms that occur intracranially. MRI findings include engorgement of dural venous sinuses, sagging of the brain, pachymeningeal enhancement, subdural effusions, and enlargement of pituitary gland.¹² Distension of the dural venous sinuses is visualized on axial and sagittal T2W images as increase in the size of flow void. Farb et al. have described this as venous distension sign.¹³ The authors have described this sign on T1WI in dominant transverse sinus on a parasagittal image (Fig. 1c). However, we feel that axial T2W images demonstrate the venous distension sign better as typical rounded flow void in the superior sagittal sinus (Figs. 1b, 7b and 9b). We think that further studies will be required to establish this observation. Sagging of brain is considered to be a very important finding, as it is the cause of most of the clinical symptoms such as diplopia (III/VI nerves), hearing changes, tinnitus, giddiness (VIII nerve), visual blurring (II nerve), and facial numbness (VII nerve) due to stretching of the cranial nerves. MRI features of descent of brain include herniation of cerebellar tonsil (Fig. 2) (which may be confused with Chiari I malformation), anteroposterior elongation of mid brain as was seen in our cases 1 and 2 (Fig. 1a and 7b), effacement of the prepontine, perisellar, and interpeduncular cisterns (Figs. 2 and 10), and low position of the iter (Fig. 8). The iter is the opening to the aqueduct of sylvius from the third ventricle which is normally 1.8–2 mm below the incisural line which connects the tuberculum sellae with the confluence of the vein of Galen and straight sinus,⁷

and the angle between straight sinus and vein of Galen becomes reduced (normal angle 49°–80°).¹⁴

Shah et al.¹⁵ evaluated objective criteria for sagging of brain, and they measured the mamillopontine distance, the pontomesencephalic angle, and the lateral ventricular or corpus callosal angle in suspected cases of IH and found that the values of mamillopontine distance of <5.5 mm and the pontomesencephalic angle of <50° were statistically significant and found that the difference in the lateral ventricular angle values between control subjects and patients with IH were not statistically significant. The mamillopontine distance was 4 mm, 4.2 mm, and 4.1 mm in cases 1, 2, and 3, respectively in our cases (Figs. 3a and 11). The pontomesencephalic angle was 33.1° (Fig. 4), 40°, and 39° in cases 1, 2, and 3, respectively in our cases.

Thick pachymeningeal enhancement involving both infra- and supratentorial compartments of brain is the hallmark of IH. This pachymeningeal enhancement is also thought to be due to venous engorgement.¹⁶ Pachymeningeal enhancement was seen in all the three cases in this case series (Figs. 5, 9 and 12). Pituitary gland enlargement has been described in IH in literature; however, we did not notice any appreciable enlargement of pituitary gland in our cases.

Subdural effusions in IH are typically thin and crescentic and may be bilateral and are usually not associated with mass effect and are thought to occur in cases where the dural venous dilatation alone is not sufficient enough to maintain the intracranial volume.¹⁷ Subdural effusion was seen in case 2 (unilateral) and case 3 (bilateral) in this case series (Figs. 7b and 10).

Spinal MR imaging is not mandatory for diagnosis of IH but recently imaging findings of spinal MRI have been included in the new diagnostic criteria of SIH.¹⁸ The spinal MRI findings include extradural CSF collections, engorged epidural venous plexus seen as flow voids on T2WI, meningeal outpouchings or diverticulae, especially along nerve root sleeves and abnormal meningeal enhancement. There are two schools of thought, one which believes that the spinal fluid collections are due to CSF leakage and accumulation and other which asserts that venous hyperemia causes transudation of fluid out of subarachnoid space resulting in spinal collections,¹⁷ which explains the high protein contents of this fluid, meningeal enhancement, and venous engorgement in these cases. Whatever may be the cause, whenever SIH is suspected, the spine should be screened with STIR sequence and if the fluid intensity is localized to a particular region that region should then be selectively imaged with CISS sequence in axial plane and attempt be made to identify the dural breach and visualize the extradural fluid collection better, as was done in case 1 (Fig. 6a and b) of this case series.

Literature suggests that CT myelography is the investigation of choice for identifying the spinal leaks in cases of SIH. Mokri et al. in their study established the site of a CSF leak in 67% of patients on CT myelography whereas spinal MRI was only able to do so in 50% of the cases.⁷ However, the CT myelography done in our case 1 could not demonstrate the CSF leak or extradural collection. This could only be explained by the fact that in our case, the opacification of the thecal sac was not uniform and circumferential and the region of dural leak remained unopacified with contrast (Fig. 6c).

Conservative management and bed rest relieve the headache. It is postulated that the CSF pressure gets reduced in supine position which helps in repair of dural defect. Other conservative methods used in treatment of IH are use of methylxanthines, theophylline, and caffeine orally or intravenously. Hydration, CO₂ inhalation, and more salt intake aim to restore CSF volume thereby preventing venous engorgement. This conservative management if given enough time is effective in majority of patients, but sometimes the symptoms may be so debilitating that specific measures may have to be taken to relieve the symptoms. Technique of epidural blood patches (EBP) for management of SIH was introduced by Gormley who observed that IH after lumbar puncture is less commonly seen after a traumatic tap or when the tap is bloody.¹⁹ In this technique, autologous blood is injected into the epidural space. Instant and long-term relief has been reported with the initial patch being effective in 85–90% of cases.²⁰ This technique can also be used even if the site of the leak is not known, the blood is injected into the epidural space in lumbar region and patient is placed in Trendelenburg position. This causes the blood to travel up into thoracic or cervical spine and block the leak due to irritant action.²¹

Surgical repair of the dural defect may be considered in patients who fail two to three EBPs, and works best in cases where a structural defect or a focal CSF leak is identified. Suturing a leaking meningeal diverticulum or a dural rent, or closing a dural hole by placement of a muscle pledget, can be performed.²¹ In our cases 1 and 3, it was decided to give a trial of conservative management before subjecting the patients to autologous epidural blood patch. Both these patients showed considerable improvement in their symptoms and were placed on sheltered appointment and on regular follow-up. In case 2, the VP shunt was readjusted to prevent over-drainage.

Intracranial hypotension as a cause of headache is uncommon but this entity should be kept in mind in patients who give history of postural headaches and in whom the headache is not relieved with routine NSAIDs. In our cases, the diagnosis of IH was delayed even though cases 1 and 3 underwent a MRI and CT head respectively. Therefore, high index of suspicion should be kept for this entity and MR examination should be carried out to rule in the diagnosis.

Conflicts of interest

The authors have none to declare.

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