INTRODUCTION

Idiopathic or Benign intracranial hypertension (BIH) is a disorder of unknown etiology characterized by raised CSF pressure. In the past, BIH was a diagnosis of exclusion and imaging played a limited role of excluding lesions producing intracranial hypertension, like “obstructive hydrocephalus, tumour, chronic meningitis, arteriovenous fistula, internal jugular vein stenosis, and dural sinus thrombosis”[1]. Of late, few subtle imaging features have been described in patients with IIH. MRI features which are seen in cases of IIH include slit like ventricles, empty sella, flattening of the posterior sclera, dilatation or tortuosity of the optic nerve sheath and gadolinium enhancement of the optic disc.

Clinical features

The typical IIH patient is an obese woman of childbearing age, with a body mass index >25. The reported incidence of IIH is 19/100,000 in middle aged females were more commonly affected. Most common radiological findings were slit like frontal horns of lateral ventricles and empty sella, present in all of our patients. Findings involving optic nerve were present in 13 out of 27 (48%) of our patients, while 2 of 27 (7%) patients had spontaneous csf leak in presence of BIH. On manometry, CSF opening pressure[OP] was more than 25cm of H2O in all the patients.

RESULTS

The study included twenty seven patients who were diagnosed with benign intracranial hypertension (BIH) based on modified Dandy’s criteria. Middle aged females were commonly affected, most common radiological findings were slit like frontal horns of lateral ventricles and empty sella, present in all of our patients. Findings involving optic nerve were present in 13 out of 27 (48%) of our patients, while 2 of 27 (7%) patients had spontaneous csf leak in presence of BIH. On manometry, CSF opening pressure[OP] was more than 25cm of H2O in all the patients.

Conclusion: Slit like ventricles, empty sella, vertical kinking of optic nerve and distension of the perioptic subarachnoid space are important radiological markers of BIH that contribute to its diagnosis.

KEYWORDS: Pseudotumor cerebri, BIH, IIH, papilledema, CSF

AIMS & OBJECTIVES:

To describe the imaging features and their frequency in benign intracranial hypertension.

MATERIALS AND METHODS:

1. STUDY DESIGN: Cross-sectional study
2. SAMPLE SIZE: 27 patients
3. INCLUSION CRITERIA:
   • All patients presented to our hospital with neurological complaints which fulfilled Modified Dandy’s criteria for BIH.

4. EXCLUSION CRITERIA:
   • Allergy to contrast agents
   • History of metallic foreign body

5. STUDY PROTOCOL:

MR imaging of the patients was performed on a Siemens 1.5 Tesla Scanner - Magnetom Essenza. After obtaining the written informed consent, MRI was performed using a dedicated head coil. After a localizer series, the standard imaging protocol consisted of the following sequences- axial T1, axial T2, axial FLAIR, sagittal T2, axial DWI and axial Gradient (Flash). Contrast (Gadodiamide) MRI was performed whenever required. Evaluation of the age, gender, symptoms, signs and opening CSF pressures was done. Various radiological findings like slit like ventricles, empty sella, flattening of the posterior sclera, dilatation or tortuosity of the optic nerve sheath and distension of peri-optic arachnoid space were documented. The frequency and distribution of each finding was calculated.

RESULTS

In our study, middle aged obese females were commonly affected, youngest patient being 20yrs & oldest being 49yrs with a mean age of 35 years.
The patients presented with headache(95%), blurring of vision(70%), retro-orbital pain(52%), pulsatile tinnitus(30%), visual loss(20%) and CSF leak(7%) in the form of CSF otorrhea.

Examination revealed papilledema, diplopia and visual loss. Radiologic examinations were performed to help exclude lesions that produce intracranial hypertension, such as tumor, chronic meningitis, obstructive hydrocephalus, dural sinus thrombosis, arteriovenous fistula etc. In our study, findings of empty sella (Fig. 1) and slit like frontal horns of bilateral lateral ventricle were found in all the 27 patients. Findings related to optic nerve such as vertical kinking of optic nerves, distension of peri-optic subarachnoid space (Fig. 2 and 3) were present in 13 out 27 patients. The CSF opening pressure in all the patients was above 25cm of H2O thus confirming the diagnosis of Benign intracranial hypertension (BIH). MRI T2W Sagittal image showing empty sella in a patient of benign intra-cranial hypertension. (BIH). Fig 1.

MRI T2W Sagittal image showing empty sella in a patient of benign intra-cranial hypertension.

**Fig 2.**  
**Fig 3.**

MRI T2W Coronal and sagittal images (Fig. 2 and 3) showing distension of the peri-optic subarachnoid space in a patient of benign intra-cranial hypertension.

**DISCUSSION**

BIH, also known as pseudotumor cerebri and idiopathic intracranial hypertension, is a syndrome characterized by increased CSF pressure and papilledema in patients without focal neurologic findings. It is a diagnosis of exclusion, and radiologic examinations are traditionally performed to help exclude lesions that produce intracranial hypertension, such as obstructive hydrocephalus, tumor, chronic meningitis, arteriovenous fistula, internal jugular vein stenosis, and dural sinus thrombosis.

Modified Dandy’s criteria is used for diagnosing IIH:  
1) If symptoms are present, they may only reflect those of generalized intracranial hypertension or papilledema.  
2) If signs are present, they may reflect only those of generalized intracranial hypertension or papilledema.  
3) Documented elevated ICP measured in the lateral decubitus position (findings are considered abnormal if above 20 cm H2O in normal-weight individuals and 25 mm H2O in obese individuals) [9].  
4) Normal CSF composition  
5) No evidence of hydrocephalus, mass, structural, or vascular lesion on MRI or contrast-enhanced CT for typical patients and on MRI and MR venography for all others  
6) No other cause of intracranial hypertension identified.

Benign intracranial hypertension (BIH) is a disorder that most commonly affects obese women of child-bearing age [1].

Vincent Giuseffi et al did a case-control study “Symptoms and disease associations in idiopathic intracranial hypertension (pseudotumor cerebri)” in which 90% of the patients were women with a mean age 33. They also found out that obesity and recent weight gain was much more common among patients than controls. Symptoms most commonly reported by IIH patients were headache (94%) and transient visual obscurations (TVO) (68%). In their study they concluded that the profile of a young obese woman with headaches and visual disturbances should alert the clinician to the diagnosis of IIH [10].

In a study by Divyata R. Hingwala et al, five imaging findings (perioptic nerve sheath distension, globe flattening, empty sella, vertical buckling of optic nerve, optic nerve head protrusion) were described in 21 patients with proven IIH and 60 patients with secondary intracranial hypertension. The patients with proven IIH had a mean age of 27.6 years (range 7-44 years) which was less than the mean age of 35 years in our study. All patients but one were females which was similar to our study.

Deterioration in vision and finally the complete loss of vision are due to pressure on the optic nerve. Obstruction of axonal transport at the level of the optic disc causes papilledema [12]. Direct transmission of the elevated CSF pressure results in distension of the perioptic subarachnoid space and ballooning of the optic papilla, causing it to protrude physically into the posterior aspect of the globe [13]. In one study, MR imaging disclosed flattening of the posterior sclera in 80%, an empty sella in 70%, distension of the perioptic subarachnoid space in 45%, enhancement of the prelaminar optic nerve in 50%, vertical tortuosity of the orbital optic nerve in 40%, and intracranial protrusion of the prelaminar optic nerve in 30% of 20 patients with IIH (14).

Eliseeva et al. in their study also noted that increased intracranial tension can be seen as prominent subarachnoid space along the optic nerve [15]. In our study findings related to optic nerve such as vertical kinking of optic nerves, distended peri-optic arachnoid space were present in 13 out 27(48%) patients. Raised intracranial pressure also leads to downward herniation of an arachnoidcele through a defect in the diaphragm sella leading to empty sella. In our study empty sella was present in all the patients.

In a study by Brodsky MC et al, the MR imaging found empty sella in 70% of patients with IIH, flattening of the posterior sclera in 80%, enhancement of the prelaminar optic nerve in 50%, distension of the perioptic subarachnoid space in 45%, vertical tortuosity of the orbital optic nerve in 40%, and intracranial protrusion of the prelaminar optic nerve in 30%. They concluded that in patients with IIH, all neuroimaging signs except for intracranial protrusion of the optic disc are highly significant for the presence of elevated intracranial pressure [16].

A retrospective study evaluating the prevalence of empty sella in patients with CSF leaks showed that 100% of patients with spontaneous CSF leak had a completely or partially empty sella turcica on imaging, compared to 11% of patients with non-spontaneous CSF leaks, and 5% to 6% of the general population without CSF leaks [17]. All patients in the spontaneous CSF leak group had elevated CSF opening pressure. Slit-like ventricles, ‘tight’ subarachnoid spaces, flattening of the posterior globe, protrusion of the optic nerve head, distension of the optic nerve sheath and vertical tortuosity of the optic nerve are other radiological findings often observed in IIH and in patients with spontaneous CSF leaks. 2 patients (7%) in our study had spontaneous CSF leak in the form of CSF otorrhea. Given the overlapping clinical and radiological presentation of primary spontaneous CSF leaks and IIH, some authors have even gone so far as to theorize that patients with primary
spontaneous CSF leaks have a variant of IIH [18]. Advances in imaging have revolutionized understanding of the intracranial pressure disturbances with improved diagnostic accuracy and patient management. Awareness of the subtle imaging findings in IIH is important to make an prompt diagnosis.

Pharmacological treatment includes acetazolamide, which is the most effective drug found to lower the intracranial pressure in these patients. For patients with inadequate headache relief with first line intracranial pressure lowering agents, primary headache prophylaxis can be started with amitriptyline, topiramate, or other commonly prescribed migraine prophylaxis agents. Corticosteroids may be used as a supplement to acetazolamide in patients who present with severe papilledema [19]. Patients presenting with progressive loss of vision should immediately be placed on oral prednisolone, if the visual field continues to worsen on corticosteroid treatment, immediate surgical management should be resorted to, which includes optic nerve fenestration surgery and Cerebrospinal fluid diversion by ventriculoperitoneal shunt or lumboperitoneal shunt [20].

CONCLUSION

BIH is a relatively less studied entity which presents with nonspecific symptoms and subtle radiological signs. Early diagnosis with prompt and adequate treatment can result in significant improvement in the clinical condition of the patient. According to our study empty sella, slit-like ventricles, vertical kinking of optic nerve and distension of peri-optic arachnoid space are important neuro-radiological markers of BIH. Thus, this study would acquaint the radiologists with various radiological manifestations of BIH which contribute maximally to its diagnosis, and hence improve the patient management.

REFERENCES