

Original article

MRI findings in Idiopathic Intracranial Hypertension

¹Dr. Bhakti Yeragi, ²Dr. Saurabh Deshpande

¹Assistant Professor, Department of Radio-diagnosis, B.Y.L. Nair Charitable Hospital, Mumbai.

²Assistant Professor, Department of Radiodiagnosis, Grant Govt. Medical College and Sir J.J. Group of Hospitals, Mumbai.

Corresponding author: Dr. Saurabh Deshpande

ABSTRACT:

Introduction-Idiopathic or Benign intracranial hypertension (IIH) presents with nonspecific symptoms and subtle radiological features which requires high degree of suspicion for diagnosis. The aim of this study is to describe clinical features, pathogenesis and radiological features of this condition.

Materials and methods-This study was carried out on 20 patients with diagnosis of BIH based on modified Dandy's criteria, between 2015-2016.

Results-Middle aged females were more commonly affected with the mean age of presentation being 39 years. Most common radiological findings were slit like frontal horns of lateral ventricles and empty sella, which were present in all our patients. Findings related to optic nerve were present in 15 out of 20 (75%) of our patients, while abnormal MR venogram was present in 13 out of 20 (65%) of patients. On manometry, CSF opening pressure was more than 25cm of H₂O in all the patients.

Conclusion: Thus slit like ventricles, empty sella, vertical kinking of optic nerve, prominence of CSF spaces around optic nerve and venous sinus abnormalities detected on MR venography are important neuro-radiological markers of IIH that contribute maximally to the diagnosis of IIH.

Keywords: Pseudotumor cerebri, papilledema, optic nerve, hypoplastic sinus, lateral ventricle

INTRODUCTION:

Idiopathic or Benign intracranial hypertension (IIH) is a disorder of unknown etiology characterized by raised CSF pressure and is primarily a diagnosis of exclusion ¹It presents with nonspecific symptoms which overlaps with many other serious neurologic disorders. Radiological signs are also subtle and a radiologist requires high index of suspicion and expertise to diagnose benign intracranial hypertension. If diagnosed appropriately it can be satisfactorily treated with good clinical improvement. Hence every radiologist must be aware of the radiological features of IIH. IIH presents with headache, nausea, vomiting, pulsatile tinnitus,

diminution of vision, double vision and other visual symptoms ². If not treated, it may progress to papilledema, which can further lead to vision loss. With this constellation of symptoms and signs, MRI forms the investigation of choice. It initially helps to exclude lesions that produce intracranial hypertension such as: Tumour, Obstructive hydrocephalus, chronic meningitis, internal jugular vein stenosis and dural sinus thrombosis, AV fistula. ³. MRI features which are seen in cases of BIH include slit like ventricles, empty sella, flattening of the posterior sclera, dilatation or tortuosity of the optic nerve sheath or gadolinium enhancement of the optic disc and dural venous sinuses abnormality.

Thus we have decided to evaluate MRI features of IIH and determine the contribution of each in reaching the diagnosis which will help the radiologist in solving this diagnostic dilemma.

AIMS & OBJECTIVES:

1) To describe the radiological features of idiopathic intracranial hypertension and the frequency and contribution of each finding for the diagnosis of this condition.

2) To describe clinical features, pathogenesis and treatment of idiopathic intracranial hypertension.

MATERIALS AND METHODS:

1. **STUDY DESIGN:** Observational prospective study

2. **SAMPLE SIZE:** 20 patients

3. INCLUSION CRITERIA:

- All cases referred to the department over a 18 month period between 2015-2016 with neurological complaints who fulfilled Modified Dandy’s criteria.

4. EXCLUSION CRITERIA:

- Patients not willing to give written informed consent.

- Allergy to contrast agents.
- Deranged renal function test (Sr.Creatinine>1.5 mg/dL)
- History of cardiac pacemaker.
- History of heart surgery/ valve replacement
- History of aneurysmal/ vascular surgery
- History of electrical implants/ neurostimulators/ pumps/ electrodes
- History of cochlear implants
- History of gunshot/ shrapnel injury/ metallic foreign body (especially in eyes).

5. STUDY PROTOCOL:

MRI imaging of the patients was performed on a “PHILIPS ACHIEVA 1.5T MRI MACHINE”, at our department. Detailed clinical histories of the patients were taken and relevant examination findings and investigations were recorded. After obtaining the written informed consent, MRI was performed using a Brain coil, according to our hospital protocol. After a localizer series, the standard imaging protocol consisted of the following sequences- axial T2, coronal FLAIR, sagittal T1, axial T2 FFE and axial DWI.

The parameters for the above sequences were-

Sequence	TE In ms	TR In ms	Matrix pixels	FOV In cm	Slice thickness In mm
T1W	15ms	Min-450ms Max-650ms	256 x	AP-215, RL-131 FH-230	5mm
T2W	100ms	shortest	512	AP-240, RL-192 FH-137	5mm
FLAIR	140ms	10000ms	288	AP-148, RL-130 FH-173	2.75

FFE	23.03	shortest	288	AP-230 RL-205 FH-149	5mm
DWI	shortest	shortest	256	AP-215 RL-220 FH-132	5mm

Evaluation of the age, gender, symptoms, signs, diagnostic evaluation(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 or gadolinium enhancement of the optic disc and dural venous sinuses abnormality were documented. The frequency and distribution of each finding was calculated.

RESULTS:

In our study, middle aged obese females were commonly affected, the mean age being 40 years, youngest patient in our study being 21yrs & oldest being 52yrs.

The patients in our study presented with headache(94%),transient visual obscurations or blurring(68%), pulsatile synchronous tinnitus(58%),pain behind the eyes(44%),double vision(38%),visual loss(30%), pain with eye movement(22%).

Examination revealed papilledema, horizontal diplopia and visual loss. Radiologic examinations are traditionally 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 slit like frontal horns of bilateral lateral ventricle (Fig.1) and empty sella (Fig.2) related to the raised intracranial pressure were found in all the

patients. Findings related to optic nerve such as vertical kinking of optic nerves (Fig.3), prominence of CSF space around optic nerves (Fig. 4) were present in 15 out the 20 patients.MR venography revealed hypoplastic left transverse sinus (Fig. 5) in 8 patients while dural venous sinuses were replaced by collaterals in 5 patients. The CSF opening pressure in all these patients was above 25cm of H₂O thus confirming the diagnosis of Idiopathic intracranial hypertension. Idiopathic intracranial hypertension cases at our institution: Table I ,II,III & IV.

DISCUSSION

Although Idiopathic intracranial hypertension (IIH), pseudotumor cerebri, and benign intracranial hypertension (BIH) are synonymous, IIH is the preferred term. Idiopathic intracranial hypertension (IIH) is a disorder that predominantly affects obese women of childbearing age.¹

Modified Dandy’s criteria is used for diagnosing IIH⁴

- 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 of

assessment of ICP by lumbar puncture are considered abnormal if above 20 cm H₂O in normal-weight individuals and 25 mm H₂O in obese individuals²⁰); MRI abnormal if above 20 cm H₂O in normal-weight individuals and 25 mm H₂O in obese individuals²⁰)

- 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.

Obesity has long been associated with IIIH. Chronic inflammation associated with obesity, raised intra-abdominal pressure leading to raised central venous pressure and increased estrogenicity associated with obesity are few factors being proposed as possible etiological factors in the development of IIIH in obese patients. Various medications and systemic diseases have been associated with IIIH^{5,6}. The most frequently reported medications are tetracyclines and vitamin A-derived medications while systemic lupus erythematosus, uraemia and hypothyroidism are the systemic diseases which have been frequently reported to be associated with IIIH⁵. Men are less commonly affected than women, are less likely to be obese and are at a twice greater risk of vision loss than women with IIIH⁷. Which part of the intracranial compartment is responsible for raising intracranial pressure in the absence of ventricular dilatation is still unclear. Many theories have been postulated which include increased venous

sinus pressure, decreased spinal fluid absorption, increased spinal fluid secretion, increased blood volume, brain oedema and idiopathic intracranial venous hypertension⁸. Sensation of fullness in the head occurs due to the compression of intracranial vasculature and cranial nerves by the increased intracranial pressure. 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 papilledema⁹. Direct transmission of the elevated CSF pressure results in distension of the perioptic subarachnoid space and also protrusion of the optic papilla into the posterior aspect of the globe¹⁰. Fixation of the distal and proximal points of the optic nerve leads to optic nerve tortuosity in patients with elevated intracranial pressure. Eliseeva et al. in their study also noted that increased intracranial tension can be seen as prominent subarachnoid space along the optic nerve¹¹. A study by Gibby et al. noted posterior globe flattening on CT scans and considered it to be the mildest in the spectrum of changes leading to the protrusion of the optic nerve head into the globe¹². Jacobson found bilateral flattening of the posterior sclera and distension of the perioptic subarachnoid space on MRI in a patient with elevated intracranial pressure and unilateral papilledema¹³. In our study findings related to optic nerve such as vertical kinking of optic nerves, prominence of CSF space around optic nerves were present in 6 out of the 8 patients. Similarly, compression of the vestibulocochlear nerve results in tinnitus and nausea. Raised intracranial pressure also leads to downward herniation of an arachnocele through a defect in the diaphragm sella leading to empty sella¹⁴. In

our study empty sella was present in all the patients. Another theory proposes that arterial CSF pulsations transmitted to the compressible medial aspects of dural venous sinuses result in periodic compression of their walls and luminal narrowing. Transverse sinus stenosis can be both a cause and an effect of intracranial hypertension. In most cases, a collapsible transverse sinus, structurally susceptible to extrinsic compression from intracranial hypertension initiates a vicious cycle. It has been postulated that raised intracranial pressure compresses the collapsible transverse sinus causing outflow obstruction, which results in venous hypertension. This then leads to decreased CSF absorption and causes further increases in intracranial pressure, which then accentuates external compression of the transverse sinus and further stenosis¹⁵. It can be seen in 14-90% patients of IIH and should be looked for in all suspected cases for IIH on MR venogram⁴. In our study abnormal MR venogram was present in 5 out of 8 (62.5%) of patients. 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 40 years in our study. All patients but one were females which was similar to our study. Of all the imaging findings evaluated, optic nerve head protrusion and globe flattening were significantly associated with IIH¹⁶. In our study

optic nerve related findings were present in 75% of the patients.

In a study by R Silbergleit et al, Magnetic resonance (MR) imaging was performed on six patients with IIH. The patients were all women with a mean age of 27 years similar to our study (range, 17-41 years). MR imaging demonstrated a partially empty sella in two patients, and one patient had dural venous sinus abnormality. Significantly larger subarachnoid space volumes were found by volumetric pixel analysis in patients than in control subjects, without a significant difference in ventricular volumes, however the wide range of normal values for the subarachnoid spaces limited the clinical use of these measurements. They also concluded that the role of MR imaging, as with CT, is primarily in the exclusion of other diseases with clinical presentations similar to that of IIH.¹⁷

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 intraocular protrusion of the prelaminar optic nerve in 30%. They concluded that in patients with IIH, all neuroimaging signs except for intraocular protrusion of the optic disc are highly significant for the presence of elevated intracranial pressure.¹⁸

Lim et al¹⁹ and Agid et al²⁰ have described the specificity and sensitivity of these imaging findings in IIH. Agid et al have shown that posterior globe flattening, optic nerve sheath distension, optic nerve tortuosity, and empty sella turcica were significantly associated with IIH and posterior globe flattening, optic nerve protrusion and slit like ventricles had

maximum specificity (100%). Lim et al found a similar high specificity for optic nerve head protrusion but lower specificity for flattening of the posterior sclera (60%). They found statistical significance only for optic nerve tortuosity. Similarly in a study by Sureyya Burcu Gorkem et al, they found out that in pediatric population, optic nerve findings and decreased pituitary gland size are reliable neuroradiological diagnostic markers for IIH²¹. Vaghela V et al concluded that although it has been many decades since the identification of this condition which presents with intracranial tension disturbances in a patient with headache, further insights into their natural history and pathogenesis are increasing. 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 diagnosis²².

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, other commonly prescribed migraine prophylaxis agents. Corticosteroids may be used as a supplement to acetazolamide in patients who present with severe papilledema²³. 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 surgery²⁴ and Cerebrospinal fluid (CSF) diversion (i.e, ventriculoperitoneal shunt or lumboperitoneal

shunt) .Intracranial venous sinus stenting within a transverse sinus stenosis is performed to decrease the pressure gradient and cerebral venous pressure. This improves CSF resorption in the venous system and leads to reduction in the intracranial pressure.^{25 26}

CONCLUSION:

IIH is a less understood and studied entity, which presents with nonspecific symptoms and subtle radiological signs.Prompt diagnosis with adequate treatment can lead to significant improvement in the clinical condition of the patient. According to our study, slit like ventricles, empty sella, vertical kinking of optic nerve, prominence of CSF spaces around optic nerve and venous sinus abnormalities detected on MR venography are important neuro-radiological markers of IIH.Thus, this study would acquaint the radiologists with various radiological markers that contribute maximally to the diagnosis of IIH and hence improve the patient management.

FIGURE 1: Axial T2 weighted MRI images shows slit like frontal horns of bilateral lateral ventricle



FIGURE 2: Coronal T2 weighted MRI images shows empty sella.

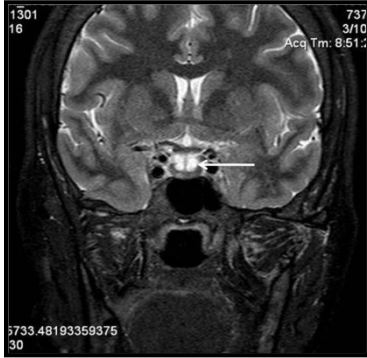


FIGURE 4: Coronal T2 weighted MRI images shows prominence of CSF spaces around bilateral optic nerves.

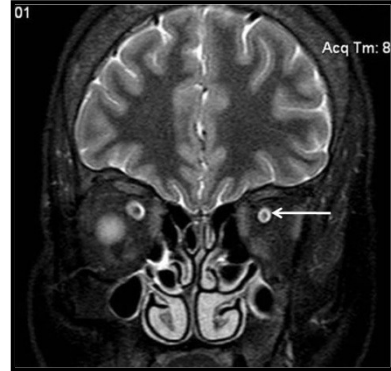


FIGURE 3: Saggital T1 weighted MRI images shows vertical kinking of optic nerve.



REFERENCES:

- 1) Michael Wall. Idiopathic Intracranial Hypertension. *Neurol Clin.* 2010 Aug; 28(3): 593–617.
- 2) S. Ambika , Deepak Arjundas , Veena Noronha, and Anshuman Ann .Clinical profile, evaluation, management and visual outcome of idiopathic intracranial hypertension in a neuro-ophthalmology clinic of a tertiary referral ophthalmic center in India.*Indian Acad Neurol.* 2010 Jan-Mar; 13(1): 37–41.
- 3) Hiroko Suzukia, Jun-ichi Takanashia, Kazuhiko Kobayashia, Kasumi Nagasawaa, Kazuyuki Tashimaa and Yoichi Kohnoa. MR Imaging of Idiopathic Intracranial Hypertension .*AJNR* 2001 22: 196-199.
- 4) A.J. Degnana and L.M. Levya. Pseudotumor Cerebri: Brief Review of Clinical Syndrome and Imaging Findings, *AJNR* 2011 32: 1986-1993.
- 5) Dhungana S, Sharrack B, Woodroffe N. Idiopathic intracranial hypertension. *Acta Neurol Scand.* 2010; 121:71–82.
- 6) Hannerz J, Ericson K The relationship between idiopathic intracranial hypertension and obesity. *Headache* 2009; 49:178–84
- 7) Bruce BB, Kedar S, Van Stavern GP, et al. Idiopathic intracranial hypertension in men. *Neurology.* 2009; 72:304–309
- 8) D Soler , T Cox P, Bullock D ,M Calver ,R O Robinson. Diagnosis and management of benign intracranial hypertension, *Arch Dis Child* 1998; 78:89-94.

- 9) Chang RO, Marshall BK, Yahyavi N, Sharma A, Huecker J, Gordon MO et al. Neuroimaging Features of Idiopathic Intracranial Hypertension Persist After Resolution of Papilloedema. *Neuroophthalmology*. 2016 May 27; 40(4):165-170.
- 10) Randy H.Kardon .The pathogenesis of visual field loss in idiopathic intracranial hypertension, *Neurology* 2008; 70:634–640.
- 11) Eliseeva NM, Serova NK, Arutiunov NV. Magnetic resonance imaging of the orbital portion of the optic nerve at different stages of papilledema. *Vestn Oftalmol* 2005; 121:5-9.
- 12) Gibby WA, Cohen MS, Goldberg HI, Sergott RC. Pseudotumor cerebri: CT findings and correlation with vision loss. *AJR Am J Roentgenol* 1993; 160:143-6.
- 13) Jacobson DM. Intracranial hypertension and the syndrome of acquired hyperopia with choroidal folds. *J Neuroophthalmol* 1995;15:178-85.
- 14) JK Agarwal, RK Sahay, SK Bhadada, Vijay Sekhar Reddy, NK Agarwal. Empty Sella Syndrome. *Journal, Indian Academy of Clinical Medicine* 2001; 2(3) 198-202
- 15) B. K. Oowler1, G. Parker, G. M. Halmagyi, I. H. Johnston, M. Besser , J. D. Pickard. Cranial Venous Outflow Obstruction and Pseudotumor Cerebri Syndrome, *Advances and Technical Standards in Neurosurgery* 2005; 30:107-74.
- 16) Divyata.R.Hingwala, Chandrasekaran Kesavadas, Bejoy Thomas, Tirur R. Kapilamoorthy, P. Sankara Sarma. Imaging signs in idiopathic intracranial hypertension: Are these signs seen in secondary intracranial hypertension too? *Ann Indian Acad Neurol*. 2013 Apr-Jun; 16(2): 229–233.
- 17) Silbergleit R, Junck L, Gebarski SS, Hatfield MK. Idiopathic intracranial hypertension (pseudotumor cerebri): MR imaging. *Radiology*.1989 Jan; 170:207-9.
- 18) Brodsky MC, Vaphiades M. Magnetic resonance imaging in pseudotumor cerebri. *Ophthalmology* 1998; 105:1686-1693.
- 19) Lim MJ, Pushparajah K, Jan W, Calver D, Lin JP. Magnetic resonance imaging changes in idiopathic intracranial hypertension in children. *J Child Neurol*. 2010; 25:294–9.
- 20) Agid R, Farb RI, Willinsky RA, Mikulis DJ, Tomlinson G. Idiopathic intracranial hypertension: The validity of cross-sectional neuroimaging signs. *Neuroradiology*.2006; 48:521–527.
- 21) Süreyya Burcu Görkem, Selim Doğanay, Mehmet Canpolat, Gonca Koc, Mehmet S. Dogan, Hüseyin Per et al. MR imaging findings in children with pseudotumor cerebri and comparison with healthy controls. *Child's Nervous System* March 2015, Volume 31, Issue 3, pp 373–380
- 22) Vaghela V, Hingwala DR, Kapilamoorthy TR, Kesavadas C, Thomas B. Spontaneous intracranial hypo and hypertensions: An imaging review. *Neurol India*.2011; 59:506–12.
- 23) Celebisoy N, Gokcay F, Sirin H, Akyurekli O. Treatment of idiopathic intracranial hypertension: topiramate vs acetazolamide, an open-label study. *Acta Neurol Scand*.2007; 116:322–327.
- 24) Chandrasekaran S, McCluskey P, Minassian D, Assaad N. Visual outcomes for optic nerve sheath fenestration in pseudotumour cerebri and related conditions. *Clin Experiment Ophthalmol*. 2006; 34:661–665
- 25) R.M. Ahmeda, M. Wilkinsonb, G.D. Parkerb, M.J. Thurtella. Transverse Sinus Stenting for Idiopathic Intracranial Hypertension: A Review of 52 Patients and of Model Predictions, *AJNR* 2011 32: 1408-1414.
- 26) Friedman DI. Cerebral venous pressure, intra-abdominal pressure, and dural venous sinus stenting in idiopathic intracranial hypertension. *J Neuroophthalmol*.2006; 26:61–64.