

Original Research Article

Clinical profile of IIH with radiological signs and correlation of these signs for diagnosis of IIH

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	International Archives of Integrated Medicine, Vol. 7, Issue 7, July, 2020.	
	Available online at http://iaimjournal.com/	
	ISSN: 2394-0026 (P)	ISSN: 2394-0034 (O)
	Received on: 08-07-2020	Accepted on: 13-07-2020
	Source of support: Nil	Conflict of interest: None declared.
How to cite this article: Veena Narisetty, Kamera Sateesh Kumar, D Eswar. Clinical profile of IIH with radiological signs and correlation of these signs for diagnosis of IIH. IAIM, 2020; 7(7): 36-41.		

Abstract

Background: Idiopathic or Benign intracranial hypertension (IIH) is a disorder of unknown etiology characterized by raised CSF pressure and is primarily a diagnosis of exclusion.

Aim and objectives: To describe the radiological features of idiopathic intracranial hypertension and the frequency and contribution of each finding for the diagnosis of this condition and correlate clinical features with the radiological findings.

Materials and methods: Prospective observational study was done in 50 patients with the diagnosis of IIH satisfying the modified Dandys criteria.

Results: Females were predominant headache and papilledema was present in all patients. Slit like frontal horns of bilateral lateral ventricle and empty sella related to the raised intracranial pressure were found in all the patients. In our study, mean CSF pressure was 315.1 mm of H₂O. Patients with Visual disturbances had higher mean CSF pressures - 327.6 mm H₂O. Mean CSF pressure in patients with VI cranial nerve palsy was 347.4 mm H₂O.

Conclusion: In our study, majority of our patients were in 2nd – 3rd decade with high female preponderance and headache is the universal presenting symptom in all patients, followed by diplopia and visual disturbances. All of them had papilledema as a predominant finding. Patients with higher CSF pressure tend to have Vision loss and VI cranial nerve palsy. In our study, slit like ventricles and empty sella was present in all the patients. 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 space around optic

nerve and venous sinus abnormalities detected on MRI are important neuroradiological markers of IIH.

Key words

Idiopathic intracranial hypertension, Headache, MRI Brain, CSF pressure.

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 [1]. It 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. 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. Aim was to evaluate MRI features of IIH and determine the frequency of each sign in predicting the diagnosis of IIH non-invasively.

Materials and methods

Prospective observational study in 50 patients with the diagnosis of IIH satisfying the modified Dandys criteria, who presented to the Neurology Department at Gandhi Medical College was done.

Demographic features, presenting symptoms, duration of symptoms, presence of any systemic disease, and relevant drug history (corticosteroids, vitamins) were be noted. All patients were subjected to focused neurological examination with particular attention to optic disc examination. The body mass index (BMI) was calculated. All patients with features of raised ICP fulfilling Modified Dandy criteria were included in the study after obtaining a written informed consent.

Inclusion criteria

- New diagnosis of IIH according to Modified Dandy criteria.
- Normal imaging (cerebral MRI and MR venogram or CT venogram normal). Documented elevation of CSF opening pressure above 25cm at lumbar puncture. Normal CSF constituents.
- Normal routine hematology and biochemistry (no abnormalities of full blood count or urea and electrolytes).

Exclusion criteria

- Intracranial hypertension due to cerebral venous sinus thrombosis, structural, metabolic or other cause.
- Other ocular disease causing visual loss.
- Systemic conditions or medications that may be associated with intracranial hypertension.

Results

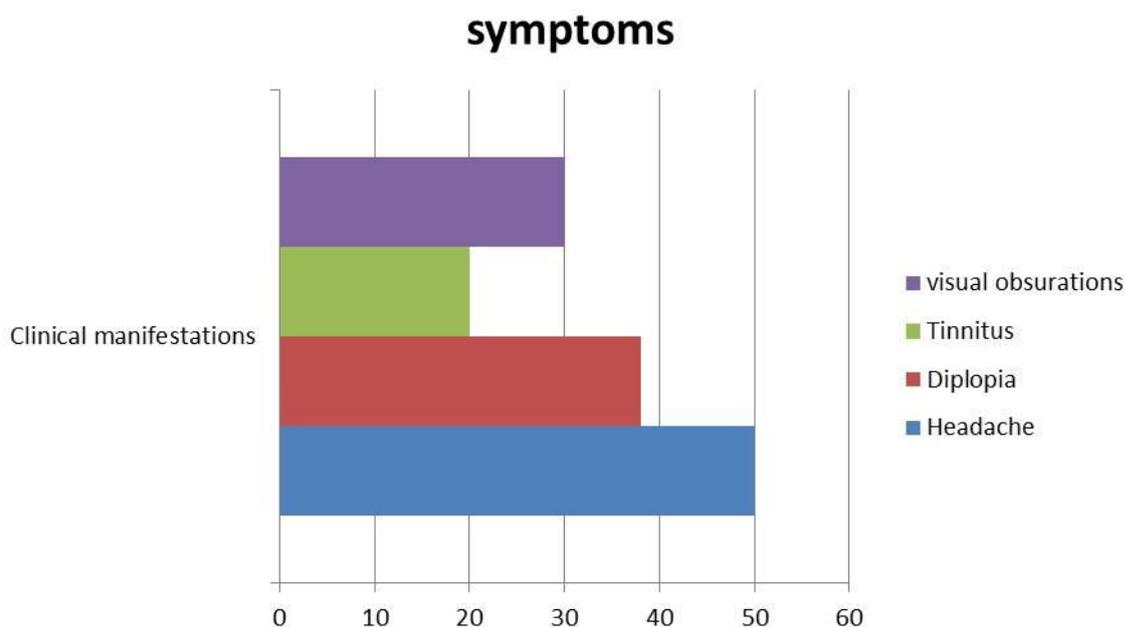
A total of 50 patients mean age was 28.12 years, 12% (n=6) were males and 88% (n=44) were females.

Symptoms like head ache, diplopia, tinnitus were analyzed among 50 patients, headache was present in all 50 patients and constituted the

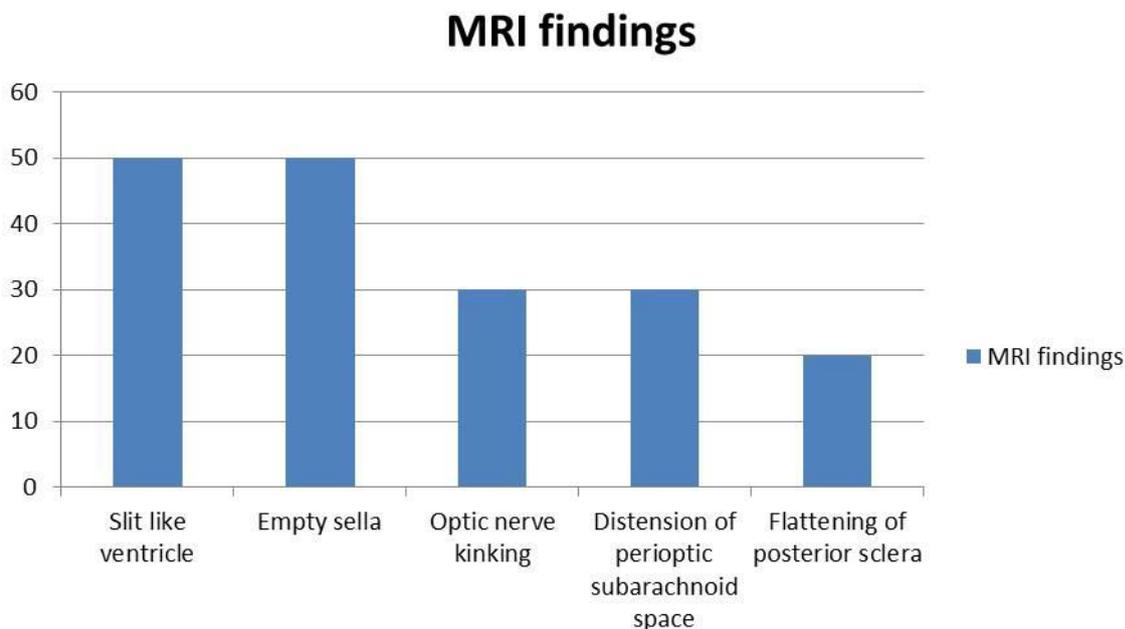
major symptoms for which patients sought neurological opinion. Diplopia was present in 38 patients. 20 patients complained of ringing sound in ears, which was unilateral in 8 patients and

bilateral in remaining. Vision disturbances in the form of visual obscurations were complained by 30 patients (**Graph – 1**).

Graph – 1: Symptoms.



Graph – 2: MRI findings.



Clinical findings

All patients underwent Neuro-ophthalmological evaluation which includes examination of fundus by Neuro-ophthalmologist and visual field charting by perimetry.

Fundus examination revealed Papilledema in all patients with bilateral Papilledema in 38 patients while 10 patients had Papilledema only in right and 4 only in left. Among 50 patients, 22 had evidence of lateral rectus palsy, of which 12 had

right, 7 patients had left and 3 patients had bilateral lateral rectus palsy.

MRI findings

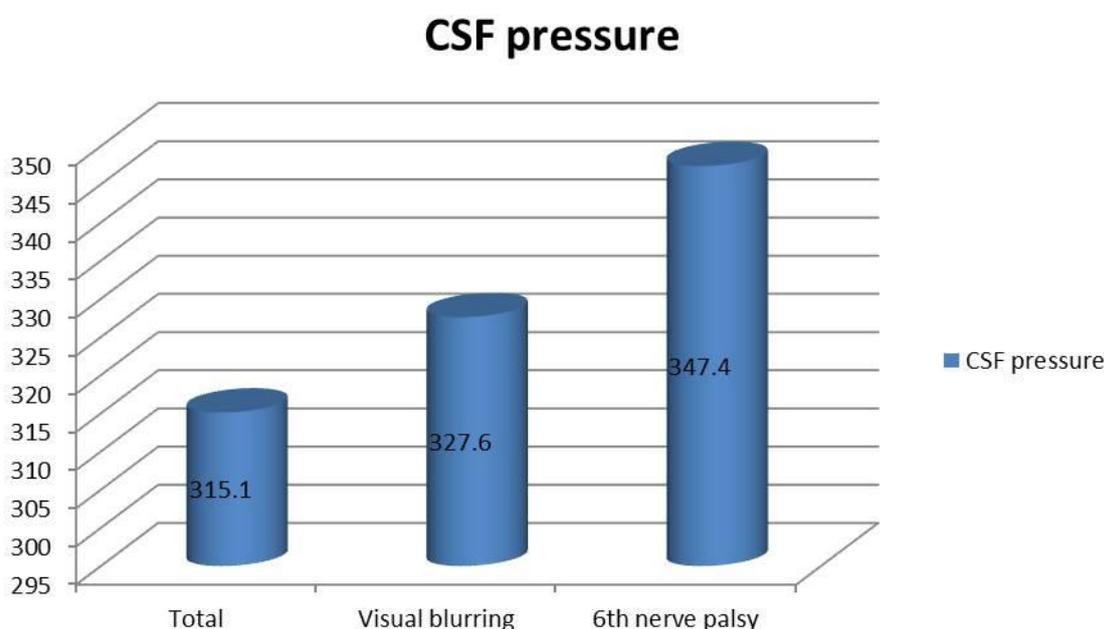
MRI brain was taken with special focus over orbital cuts MRI changes including slit like ventricle, empty sella, optic nerve kinking, distension of perioptic subarachnoid space, flattening of posterior sclera were noted. In our study, out of 50 patients findings of slit like frontal horns of bilateral lateral ventricle and empty sella related to the raised intracranial pressure were found in all the patients. Findings

related to optic nerve such as vertical kinking of optic nerves prominence of CSF space around optic nerves were present in 20 patients and flattening of posterior sclera in 20 patients (**Graph – 2**).

CSF analysis

All 50 patients underwent lumbar puncture and CSF analysis including biochemical, microbiological, cytology and pressure studies were done after obtaining written consent and under strict aseptic precautions (**Graph – 3**).

Graph – 3: CSF pressure.



All patients had normal biochemical values (sugar, protein, sodium + chloride). All of them were acellular. CSF Opening pressure greater than 250 mm of CSF was taken as abnormal.

Mean CSF pressure was 315.1 mm H₂O. Patients with Visual disturbances had higher mean CSF pressures - 327.6 mm H₂O. Mean CSF pressure in patients with VI cranial nerve palsy was 347.4 mm H₂O.

Discussion

The pathophysiology of this condition is still unclear. Due to greater awareness of this

condition, this condition is now being diagnosed with increasing frequency. It is crucial to differentiate between idiopathic and secondary causes of raised intracranial tension [4]. Severe IIH may lead to visual deterioration, which can be prevented by optic nerve sheath fenestration. Appropriate treatment can be instituted for secondary causes like venous sinus thrombosis or meningitis.

Age

The Youngest age in our study was 18 and the oldest was 50 years old, with mean age of 28.12 years. This correlates slightly lower with study

by Wall and George (1991) [5] in which mean age was 31 years. IIH is a disease mostly affecting females and there is a clear female preponderance in previous studies done by Radhakrishnan et al. [6]. This study consisted of 88% Females in accordance with previous studies.

Symptoms

Headache was present in all 50 Patients in our study, Headache has a quality of raised ICP in all of them, and majority experience vomiting and nausea at least once during this course of illness.

Various studies like Wall and George (1991) [5], Radhakrishnan, et al. (1993) [6], Kesler, et al. [7] all noted a similar prevalence of headache among IIH patients (80-98%). Diplopia was noted in 38 (76%) of our patients and most of them described it as double vision for distant objects and on looking to either side. Visual obscurations was noted in 30 (60%) of patients in our study. Similar incidence was noted in various other studies between 57-72% [5, 6].

Clinical Signs

Papilledema is almost a universal finding in IIH patients all patients had papilledema either unilateral or bilateral. Bilateral papilledema was noted in 36 patients whereas right alone in 10 patients and left eye alone in 4 patients.

In our study, we found VIth cranial nerve palsy in 22 patients (44%). Lateral rectus palsy was found in 6 (42.8%) cases in study by Ayush Dubey [8] on Idiopathic Intracranial Hypertension (IIH) in a tertiary referral teaching centre in Central India.

Visual field defects

The most common field defect noted is enlargement of blind spot, and this finding correlated with those patients who had papilledema.

CSF pressures

In our study, mean CSF pressure was 315.1 mm of H₂O. Patients with Visual disturbances had

higher mean CSF pressures - 327.6 mm H₂O. Mean CSF pressure in patients with VI cranial nerve palsy was 347.4 mm H₂O.

Imaging

In our study, slit like ventricles and empty sella was present in all the patients. Optic nerve kinking and distension of perioptic subarachnoid space was present in 30 out of 50 patients and flattening of posterior sclera in 20 out of 50 patients.

Surgery

Surgery was done in 7 patient enrolled in our study, who had recurrence of symptoms at the end of three month treatment period. Worsening of vision was the indication for surgery in all the 7 patients. All of them underwent lumbo peritoneal shunt.

Conclusion

In our study, we analyzed the demographic features, clinical presentation, investigations and outcome of IIH patients, following conclusions were observed. Majority of our patients were in 2nd – 3rd decade. There was high female preponderance in our study. Headache was the universal presenting symptom in all patients, followed by diplopia and visual disturbances. All of them had papilledema as a predominant finding, followed by lateral rectus palsy but no other focal neurologic deficits. Patients with higher CSF pressure tend to have Vision loss and VI cranial nerve palsy. In our study, slit like ventricles and empty sella was present in all the patients. Optic nerve kinking and distension of perioptic subarachnoid space was present in 30 out of 50 patients and flattening of posterior sclera in 20 out of 50 patients. IIH was 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 space around optic

nerve and venous sinus abnormalities detected on MRI were important neuroradiological markers of IHH. Thus, this study would acquaint the radiologists with various radiological markers that contribute maximally to the diagnosis of IHH and hence improve the patient management. Surgical management is essential in patients with Vision loss or impending vision loss.

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