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STUDY OF CLINICAL SPECTRUM OF IDIOPATHIC INTRACRANIAL HYPERTENSION IN A TERTIARY CARE CENTER IN SOUTH INDIA

Neurology

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ABSTRACT

Context: Idiopathic intracranial hypertension (IIH) predominantly affects young, obese women.[1] Main clinical presentations are headache, vomiting and visual symptoms. IIH is diagnosed in accordance with Modified Dandy's Criteria.The revised diagnostic criteria for IIH was proposed by Friedman et al in 2013 [3] which defines IIH with and without papilledema. If untreated IIH can progress to vision loss.[2] **Aims:** We aimed at evaluating the clinical and radiological spectrum of patients diagnosed as Idiopathic Intracranial hypertension (IIH) in a South Indian tertiary care centre. **Settings And Design:** This was a cross-sectional study conducted in patients presenting to Department of Neurology, Thanjavur Medical College Hospital. **Methods And Material:** A total 22 adult patients who fulfilled the revised diagnostic criteria for IIH proposed by Friedman et al in 2013 were included in this study. Detailed history, clinical examination and investigations including CSF opening pressure, MRI Brain with MR venogram of each patient were recorded and analysed. **Results:** Obesity and female gender are an important risk factors for IIH, but atypical IIH in the form of non-obese patients, male gender was observed in our study. Unusual findings like anosmia, fulminant IIH with third nerve, seventh nerve involvement, IIH with CSF leakage and IIH without papilledema were seen in our study. Most common MRI abnormality observed was widening of peri optic space followed by partial empty sella. **Conclusions:** Improved understanding of the clinical profile and neuroimaging of IIH can help to prevent diagnostic delay and promote timely intervention to preserve vision.

KEYWORDS

Idiopathic intracranial hypertension, headache, obesity, papilledema, neurology

INTRODUCTION:

Idiopathic intracranial hypertension is a disease characterised by raised intracranial pressure that predominantly affects young, obese women.[1] Idiopathic intracranial hypertension (IIH) was earlier known as benign intracranial hypertension (BIH) or pseudotumorcerebri (PTC). The main clinical presentations are headache, nausea, vomiting, pulsatile tinnitus, double vision, and other visual symptoms. Fundus abnormalities include papilledema of varying grades. If untreated IIH can progress to vision loss.[2]

IIH is diagnosed in accordance with Modified Dandy's Criteria.The Revised Diagnostic criteria for IIH was proposed by Friedman et al in 2013 [3] which defines IIH with and without papilledema. According to this, IIH Diagnostic criteria includes papilledema, normal neurological examination except 6th cranial nerve palsy, neuroimaging evidence for normal brain parenchyma (no hydrocephalus, mass, structural lesion or meningeal enhancement) with venous thrombosis excluded in all, normal CSF constituents and elevated lumbar puncture pressure >25 cm CSF. IIH without papilledema (IIHWOP) is diagnosed when criteria for IIH except CSF pressure fulfilled plus unilateral or bilateral sixth cranial nerve palsy. This criterion highlights the importance of a pressure greater than 25cm of CSF and the necessity for additional features such as sixth nerve palsy and MRI imaging features, which suggest pathologically raised ICP.

Various subtypes of IIH have been described. Typical IIH is that which occurs in patients who are female, of childbearing age and who have a body mass index (BMI) greater than 30.[4] Atypical IIH is the one in patients who are not female, or not of childbearing age or who have a BMI below 30. These patients require more in-depth investigation to ensure no other underlying causes. Fulminant IIH refers to patients with precipitous decline in visual function within 4 weeks of diagnosis of IIH. [5-7] IIH without papilledema is a rare subtype of IIH and is seen in patients who meet all the criteria of definite IIH in the absence of papilledema. [3]

Typical neuroimaging features of raised intracranial pressure include empty sella, partially empty sella, increased tortuosity of optic nerve, enlarged optic nerve sheath, flattened posterior sclera, intraocular protrusion of optic nerve head and attenuation of the cerebral venous sinuses including bilateral transverse sinus stenosis or stenosis of a dominant transverse sinus.[8]

The main principles of management of IIH are to treat the underlying disease, to protect vision and to minimise headache and morbidity. Weight loss is the only disease-modifying therapy in typical IIH.[9]

There are only few studies in India on Idiopathic Intracranial Hypertension (IIH) based on the Friedmans revised 2013 criteria which explore clinical spectrum of IIH including, rare, atypical presentations, and fulminant types. We studied IIH patients presenting to a tertiary care centre in South India and their clinicoradiological profile.

Subjects And Methods:

This was a cross sectional study conducted in patients presenting to Department of Neurology, Thanjavur Medical College. The aim of the study was to evaluate the clinical profile, CSF pressure and imaging findings of patients with Idiopathic Intracranial hypertension (IIH). Patients who fulfilled the revised diagnostic criteria for IIH proposed by Friedman et al in 2013 were included in the study. Patients with Intracranial hypertension due to cerebral venous sinus thrombosis, structural, metabolic, or other causes were excluded from the study. A total of 22 patients were included in this study .

History:

Detailed history regarding symptoms including headache and its duration, severity, visual symptoms , additional symptoms if any were noted. History regarding possible risk factors were also taken.

Clinical Examination: A complete neurological examination was performed. Best corrected visual acuity was recorded. Perimetry and fundus photography was also taken.

Obesity Assessment: Height, weight was noted and body mass index (BMI) was calculated.

Investigations: MRI Brain including MR venogram and CSF opening pressure using a CSF Manometer was recorded.

RESULTS:

We studied 22 cases of patients with IIH and analysed the epidemiological profile, symptomatology, clinical and radiological profile of each of these patients. This is depicted in table 1,2, 3 and 4 respectively. Of the 22 patients included in the study, 20 patients (90.9%) were female and 2 patients (9%) were male. Age group of the patients ranged from 18 to 47 of which, 1 patient in 18-20 years, 6 patients 21-29 years, 13 patients belonged to 31-40 years and 2 patients > 40 years of age. Mean age was 32.75 years.

Out of the 22 patients, 19 patients did not have any co morbidities ,3 patients had history of taking hormonal medications. Most common presenting symptom was headache, other symptoms were blurring of vision, double vision and few patients had rare symptoms like facial weakness, anosmia. Headache was a presenting symptom in all our patients. Visual symptoms were present in 14 patients and ranged from

transient blurring of vision to unilateral or bilateral visual diminution or vision loss. 7 patients had diplopia.2 patients had tinnitus, 2 patients had Facial weakness, 1 patient had CSF leakage from nose. Duration of symptoms at presentation was variable ranging from years to days. Longer duration of headache was noted whereas visual symptoms lasted weeks to days and brought patient to hospital.

Of the 22 patients, 10 patients had headache for more than 1 year duration, 9 patients had more than 1 month history of headache and 3 patients had less than 2 weeks headache duration. It was the sole presenting symptom in 4 patients. Headache was bilateral in 12 patients, unilateral in 10 patients. It was persistent throbbing type moderate to severe intensity headache in majority. 2 patients with headache for more than 4 year duration, had migraine like headache associated with nausea and vomiting.

14 patients (63.63%) had blurring of vision of which 2 patients had transient blurring of vision. Visual symptoms were bilateral in 11 patients, while 3 patients had unilateral symptoms. Only 2 patients had vision loss. Diplopia was one of the presenting symptoms in 8 patients and duration ranged from 4 months to 2 days. Pulsatile tinnitus was present in 2 patients which was unilateral and of more than 1 year duration. 2 of our patients also had anosmia, one among them had leakage of CSF from nose of 3 month duration. Facial weakness was present in 2 patients, both of whom had severe vision loss as well.

We came across both obese and non-obese patients.9 patients were obese (40.49%),4 were overweight (18.18%) and rest had normal BMI (36.36%). Mean BMI was 27.049. Most of our patients had normal vision or mild visual impairment (visual acuity >/= 20/70). 6 patients had normal visual acuity, 5 had mild visual impairment. 8 patients had moderate visual impairment (VA >/= 20/200). 3 patients had severe visual impairment, 2 of them had only perception of light in both eyes. Visual impairment was unilateral in one patient, rest had bilateral involvement. Visual field when assessed clinically was normal for most of the patients with mild to moderate visual impairment, 7 were noted to have enlarged blind spot on clinical examination.10 patients (45.45%) on examination had abducens nerve palsy which was bilateral in 4 patients and unilateral in 8 patients. One patient had third nerve palsy along with 6th nerve palsy. LMN seventh nerve palsy was noted in 2 patients and 2 patients on examination had anosmia.

Fundus examination in our patients revealed grade 4 papilledema in 6 patients, grade 3 in 4 patients and grade 2 in 6 patients. 2 patients had grade 1 papilledema, while 2 patients had mild disc pallor as the fundus finding.2 patients did not have papilledema. It was noted that the grade of papilledema correlated with the severity of visual impairment, CSF opening pressure ranged from 250 mm of CSF to 640 mm of CSF.13 patients (59.09%) had CSF pressure measured between 250 mm and 300 mm, 9 (40.9%) had pressure above 300 mm of CSF. Mean CSF pressure was 353.18 mm of CSF. BMI did not show correlation with severity of visual impairment or CSF opening pressure.

In our study MRI brain including MR venogram did not reveal any abnormal findings in 6 patients. Most common abnormalities seen were widening of peri-optic CSF space, partial empty sella and tortuosity of optic nerve. Figure 1 shows MRI images from our study. 12 patients had partial empty sella and 2 had empty sella. Bilateral optic nerve tortuosity was present in 10 patients and widening of peri optic space noted in 13 patients. Flattened posterior sclera noted in 3 patients of whom one had intraocular protrusion of optic nerve. This correlated with severe IIH. Slit like ventricles noted in 2 patients.MR venogram was normal in most of the patients, except 2 patients who had left transverse sinus stenosis.

Of the 22 patients included in our study, 18 patients were diagnosed as IIH by 2013 diagnostic criteria and 4 fulfilled IIH without papilledema criteria. Fulminant IIH was seen in 2 patients and IIH with CSF Rhinorrhoea in one patient.

The 2 patients with Fulminant IIH were female and obese. They had severe headache and vomiting. Both had severe vision impairment, ophthalmoplegia including third nerve involvement in one patient. Seventh nerve involvement was present in both of them, and both had grade 4 papilledema. CSF pressure was well above 300 mm of CSF. MRI showed empty sella, flattening of posterior sclera and optic nerve tortuosity. Both the patients underwent surgical procedures.

IIH without papilledema was diagnosed in 4 patients of whom 2 had normal disc and 2 had mild pallor. These patients had sixth nerve involvement and imaging findings fitting into Friedman's Criteria.

18 patients were discharged with improvement in symptoms, 2 patients underwent LP shunt, one patient underwent optic nerve sheath fenestration surgery. Patient with CSF rhinorrhoea had closure of the defect and and LP shunt.

Table 1 : Epidemiological Profile

Patient characteristics	Number	%
Total	22	
Gender		
Female	20	90.9 %
Male	2	9 %
Age distribution		
18-20	1	4.5%
21-30	6	27.2 %
31-40	13	59 %
>41	2	9%
(mean age :32.7)		
Risk factors		
none	19	86.36%
hormonal contraceptive	3	13.63 %
BMI		
normal	9	40.9%
overweight	4	18.18%
obese	8	36.36 %
(mean : 27.049)		

Table 2 : Symptomatology

Symptom	No	%
Headache	22	100 %
unilateral	12	54.5%
bilateral	10	45.4 %
Duration		
> 1 year	10	
2-12 months	9	
< 2 months	3	
Blurring of vision	14	63.63 %
unilateral	3	
bilateral	11	
visual symptom		
transient	2	9 %
blurring	10	45.45 %
vision loss	2	9 %
Double vision	8	36.36 %
Tinnitus	2	9%
Unilateral Facial deviation	2	9 %
Anosmia	2	9 %
Drooping of eyelid	1	4.5%
CSF leakage from nose	1	4.5 %

Table 3 : Clinical Profile

Clinical findings	No.	%
Vision impairment		
normal	6	27.27 %
mild (>/= 20/70).	5	22.72 %
moderate (>/= 20/200).	8	36.36%
severe	3	13.63 %
Papilledema		
Grade 1	2	9%
Grade 2	6	27.27%
Grade 3	4	18.18%
Grade 4	6	27.27%
None	2	9%
Disc pallor	2	9%
Cranial nerve		
anosmia	1	4.5%
3rd nerve	1	4.5%
6th nerve	10	45.45%
bilateral	4	
unilateral	6	
7th nerve	2	9%
CSF opening pressure		
>300 mm	9	40.9%
250-300	13	59.09%

Table 4 : Radiological Profile

MRI Brain Finding	No	%
Normal imaging	6	27.2%
Partial empty sella	12	54.5%
Empty sella	2	9%
Widening of perioptic CSF space	13	59%
Tortous optic nerve	10	45.4%
Slit like ventricles	2	9%
Flattened posterior sclera	3	13.6%
Intraocular protrusion of optic nerve	1	4.5%
Transverse sinus stenosis	2	9%

Figure 1: Neuroimaging Findings

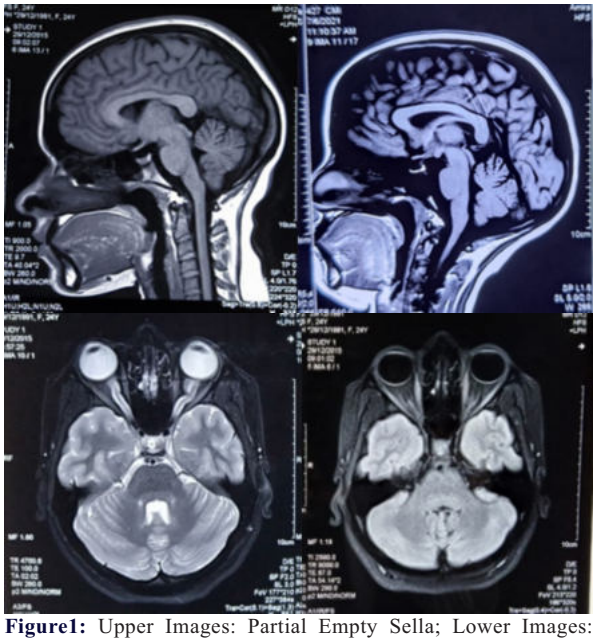

Figure1: Upper Images: Partial Empty Sella; Lower Images: Widening Of Perioptic CSF Space And Tortous Optic Nerve

DISCUSSION:

IIH is classically described to occur in obese females of childbearing age group. Majority of the patients in our study were females, and belonged to age group between 31-40 years. In this study 40.9 % patients had obesity. However it is notable that 9% patients were male and 36.36% of patients had a normal BMI.Both male patients had normal BMI. In this study, BMI did not correlate with the severity of the disease.

A study by Pal A et al in Indian patients using Friedmans criteria evaluated 33 patients (31 female and 2 male) of which 25 patients had BMI less than 25. [10]

Clinical presentation of our patients were similar to various previously published studies.[10-12] In a case-control study, the symptoms most frequently reported by IIH patients were headache (94%), transient visual obscuration (68%), pulsatile tinnitus (58%), photopsia (54%), diplopia (38%), vision loss (30%), and retrobulbar pain on eye movements (22%),[13] Headache was the commonest symptom and was present in all our patients.

Most of our patient had normal vision or mild visual impairment. Transient visual symptoms were noted in 2 patients. Both Male patients in our study were noted to have severe visual impairment. Fulminant IIH patients had very severe and rapid vision impairment. In our study 9% patients had vision loss. In a study of 43 female patients by Baheti et al also, only 9% of patients had vision loss at presentation.[14]

We also had patients with unusual symptoms like facial weakness, third nerve involvement, anosmia, CSF rhinorrhoea which are not commonly reported in other studies. In particular, both patients with facial weakness had fulminant IIH at presentation requiring surgical interventions. Third nerve palsy was also seen in one of the fulminant IIH patient.

Papilledema is considered an important sign in IIH.Our patients had varying grades of bilateral papilledema and grade was seen to correlate with clinical severity. Visual impairment was severe in those with other cranial nerve findings and severe grade of papilledema. Among our patients,4 (18%) were diagnosed as IIH without papilledema. In a study by Digre et al in 353 patients with IIH, prevalence of IIH without papilledema was 5.7%,[15]

Abducens nerve palsy, either unilateral or bilateral is reported in about one-third of patients.[16] In our study 45.45 % of patients had abducens nerve involvement either bilateral or unilateral. 1 patient had unilateral third nerve involvement as well. CSF opening pressure was very high (>300 mm of CSF) in 40.9% patients and correlated with severity of IIH.

27.2 percent of patients had a normal MRI imaging. Most common abnormality noted was widening of perioptic optic space (59%) followed by partial empty sella (54.5%) and optic nerve tortuosity (45.4%). 9% patients had slit like ventricles. This is similar to few previous studies on imaging findings in IIH. In a study by Sultan et al optic hydropes and empty sella turcica were the most common MRI abnormalities occurring in 95.8% and 70.8%, respectively.[12]. In a study on Indian population by Pal et al most common MRI finding was flattening of posterior aspect of globe and was found in 90.90%.[10].

There are only few studies based on the 2013 modified criteria for IIH in the Indian population. Our study had different spectrums of IIH patients namely typical IIH, atypical IIH (non obese patients, male IIH), fulminant IIH and IIH without papillodema. Most of the patients responded to treatment and were symptomatically better at discharge.

CONCLUSION

Obesity and female gender are important risk factors for IIH, but Atypical IIH in the form of non obese patients, male gender was observed in our study. Headache is the most common symptom. Visual impairment ranges from mild to severe vision loss. Unusual findings like third nerve involvement,seventh nerve involvement and CSF leakage were seen in our study.

IIH without papilledema can present with similar symptoms. In our study, grade of papilledema and CSF pressure correlated with the severity of symptoms. MRI was found to be normal in one third of IIH patients. Most common MRI abnormality observed was widening of peri optic space followed by partial empty sella.

Fulminant IIH patients in this study invariably had severe visual impairment as well as uncommon findings like other cranial nerve involvement. It is important to recognise the features of Fulminant IIH and make diagnosis early as these patients may require surgical interventions to save vision.

The limitation of this study is perhaps the cross-sectional nature and the small subpopulation involved. More research may help expand the understanding of the clinical spectrum of IIH ranging from mild to fulminant. Being a cause for preventable visual loss, prompt and better clinical recognition of IIH is of paramount importance to help prevent diagnostic delay and timely interventions.

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