Neurology Asia 2023; 28(4) : 999 – 1008

Clinical, radiological profile and outcomes of patients with idiopathic intracranial hypertension: a prospective study in the South Indian population

Mohamed Azharudeen *DM Neurology*, Ravi Lalapet Anbazhagan *DM Neurology*, Kannan Vellaichamy *DM Neurology*, Chandramouleeswaran Venkatraman *DM Neurology*, Balasubramanian Samivel *DM Neurology FRCP*

Institute of Neurology, Madras Medical College, Chennai, India

Abstract

Objective: To describe the clinical, radiological characteristics and outcomes of patients with Idiopathic intracranial hypertension (IIH) at 6 months in a cohort of South Indian patients. *Methods:* We did a prospective study on patients presented to our institute with the symptomatology of IIH. A detailed history regarding headache, visual symptoms were noted. A complete neurological examination including examination of the fundus, CSF opening pressure, MRI brain, and MR cerebral venography were performed for all subjects. The patients were followed-up for 6 months to assess treatment response. *Results:* We evaluated 49 patients, the majority were females, and 38.7% had grade 1 obesity. Headache was the most common clinical presentation. The most common MRI abnormality was empty sella turcica, noted in 47%. There was a significant positive correlation between CSF opening pressure and headache severity (R2= 0.165, p = 0.018), as well as between CSF opening pressure and the grade of papilledema (R2=0.245, p=0.001). Of the cases, 95% responded to medical management, and three patients of fulminant IIH underwent lumbar-peritoneal shunting. At 6 months, 60% had persistent headaches. Only 8.7% had persistent visual symptoms, and only one had optic atrophy. There was no significant correlation between the follow-up grade of papilledema and the patient's BMI. No significant correlation was found between the amount of weight lost and the improvement in the Numerical Rating Scale (NRS) score at 6 months.
Conclusion: Headache in IIH is multifactorial and a significant cause of disability. The overall prognosis for vision is good. IIH should be suspected in any obese woman with headaches since it is one of the preventable causes of blindness.

Keywords: Idiopathic intracranial hypertension, obesity, headache, papilledema

INTRODUCTION

Idiopathic intracranial hypertension (IIH) is a disorder of elevated intracranial pressure (ICP) in the absence of overt brain lesions and with normal cerebrospinal fluid (CSF) composition.[1] The rise in the prevalence of obesity is an emerging reason for the increase in the number of IIH cases in the Indian population. Children, men, and non-obese individuals may also be affected. The prevalence is high, taking into account the chronic nature of the disease. Its pathophysiology is still debated. Several theories have been proposed including increased CSF outflow resistance, venous microthrombi, fat metabolism, abnormal vitamin A metabolism, endocrine deregulation, and altered sodium and water levels in the brain.[2] The most dreadful complication of IIH is loss of vision, which can be inevitable if not treated immediately. The vision loss may occur early or late in the disease process.[3] The course of IIH can vary from a short, self-limiting illness to more severe syndromes that result in rapid vision loss.[4] Few large-scale studies in Asia have analyzed the long-term outcome of IIH in terms of recurrence rate/visual function. The goal of this research is to analyze the current clinical and radiological features of IIH patients and assess their long-term outcomes in a cohort of South Indian patients.

Address correspondence to: Mohamed Azharudeen, Institute of Neurology, Madras medical college, Chennai- 600001, India. Tel: -8526587272, email: azharudeen1226@gmail.com

Date of Submission: 2 October 2023; Date of Acceptance: 7 October 2023

https://doi.org/10.54029/2023ijr

METHODS

This was a prospective cohort study conducted in the Institute of Neurology, Madras Medical College, which is a tertiary referral center in South India during the period 2021-2023. Patients fulfilling the modified dandy criteria for IIH[5] were recruited. Demographic details such as age, gender, and body mass index (BMI) were documented. BMI is calculated as weight in kilograms divided by height in meters squared (kg/m2) and is categorized according to the WHO, as underweight (<18.5), normal weight (18.5 to 24.9), overweight (25 to 29.9), and obesity class 1(30 to 34.9), obesity class 2 (35to 39.9), obesity class 3(>40).[6] All patients were subjected to a thorough medical history, including the onset, duration, frequency, location, nature, severity, and type of headache. A manual review of each participant's headache symptoms was performed to define the headache phenotype, which was then categorized according to the International Classification of Headache Disorders (ICHD-3). To measure the intensity of the headache, a Numerical Rating Scale (NRS) was employed. Patients rated headache severity on a scale from 0 to 10. Patients with scores of 1 to 3 were considered to have mild headaches, 4 to 6 were deemed to have moderate headaches, and 7 to 10 were deemed to have severe headaches.[7] Visual complaints (such as visual obscurations, blurred vision, diplopia, and visual loss) as well as accompanying symptoms including nausea, vomiting, photophobia, phonophobia, and tinnitus were also noted. Past medication histories were also reviewed with an emphasis on systemic corticosteroids and oral contraceptive pills. A complete neurological examination was performed for all patients with a special focus on anterior segment examination, visual acuity, extraocular movements, pupil evaluation, color vision, confrontation test for visual field defects, measurement of intraocular pressure, and fundus examination with indirect ophthalmoscopy. Humphrey's 30-2 visual field analysis was performed. Fundus examination was done to determine the grade of papilledema using a modified Frisén scale. A lumbar puncture was performed to measure the opening pressure of cerebrospinal fluid in the lateral decubitus position with the patient's legs relaxed and slightly extended, using a manometer. CSF analysis was done to rule out any abnormality. To rule out structural causes of elevated intracranial pressure and cerebral venous thrombosis, MRI brain with contrast and MRV was performed. Depending on the severity of symptoms, patients were treated with either a drug therapy (eg, acetazolamide, topiramate, or both) or a combination of drug therapy with ventriculoperitoneal shunting. Patients were followed up for 6 months, which included two visits, one at the third month and the other at 6 months. Patients were asked to maintain a headache diary which contains details of headache duration, frequency, severity, and analgesic use (No of days used). The headache diary is reviewed during each visit; in addition, details regarding visual symptoms and fundus examination were noted. Visual field analysis by automated perimetry was done to assess the treatment response.

Statistical analysis

The qualitative variables were displayed as frequencies and percentages, while the quantitative data were represented as means and standard deviations. The paired t-test was used to compare continuous data in the research group. An independent t-test was used to compare continuous variables between two groups, while one-way ANOVA was used to compare more than two groups. Chi-squared test was adopted for group comparisons for categorical data. For correlation between two normally distributed quantitative variables, the Pearson coefficient was utilized. Data was analyzed using SPSS v23 (IBM Corp.) software. All statistical analysis was carried out at a 5% level of significance and p-value <0.05 was considered significant

Ethic

All patients gave written informed consent before participating in the study. The ethics committee of the institute approved the study protocol.

RESULTS

Demographic data

Among 49 subjects, the majority were females (98%). The age was in range from 16 to 62 years, mean ± SD age of the cases was 29.8 ± 9.7 years. Forty-nine percent of cases were in the age group of 20-30. Age-wise distribution of our study population is depicted in Figure 1. The distribution of BMI in the study population is shown in Table 1. The majority (38.7%) of cases were overweight. The mean ± SD of BMI was 31.5 ± 5.8 kg/m2.

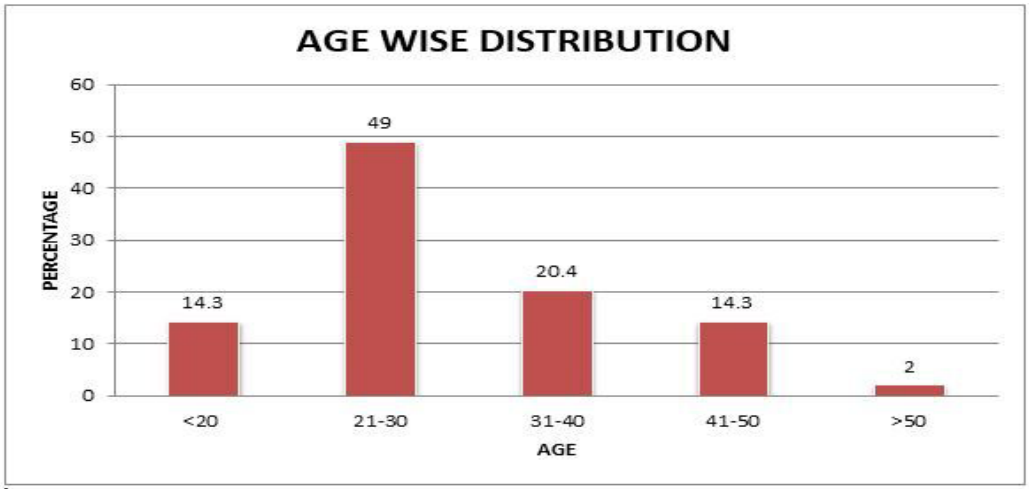

Figure 1. Age wise distribution of the study population.

Clinical features at the presentation

Headache was the most frequent complaint in the study participants followed by transient visual obscuration and visual impairment [Table 2]. All patients had headaches (100%), and the duration of headaches ranged from 25 days to 3 years. Among these cases, 23 (46.9%) had continuous headaches that occurred daily. Twenty patients (40.8%) experienced intermittent chronic headaches (more than 15 headache days per month), and 6 (12.2%) had episodic headaches that occurred less than 15 days per month. Four subjects (8%) presented with bi-temporal headache, global headache in 11 cases (22.4%), 12 patients (24.5%) presented with unilateral temporal headache, occipital headache in 9 cases (18.4%), and occipito frontal headache in 13 cases (26.5%). The most frequent headache type reported was throbbing (30%) followed by a dull ache in 28%. The headache phenotypes described are a migraine-like headache in 68%, 39 % had migraine-like without aura and 29% had migraine-like with aura, tension-type headache phenotype in 20% of subjects, and medication overuse headache in 7%, and in rest, headache could not be classified into a particular category.

The majority of the patients (44%) reported the severity of headaches as 4-6 on the numerical rating scale (NRS). Twenty patients (40%) reported severe headaches and 8 cases (16%) had mild headaches. The severity of headache positively correlated with CSF opening pressure before treatment (R^2= 0.165, p = 0.018) as presented in Figure 2. Transient visual obscuration was the second most common symptom in our study cohort. A subset of patients (24.5%) had radicular pain in the neck and shoulder, and back pain. This radicular pain is hypothesized to occur when spinal dural root sheaths are filled by CSF under high pressure. The median time from symptom onset to diagnosis was 70 days (range: 30-1080 days). Only 15 sought immediate medical advice, while 34 patients (69%) had multiple reasons for a delay in diagnosis. The most common reason was symptom neglect in 12 (35%) patients. It is often misdiagnosed as migraine, tension-type headache, sinusitis, medication overuse headache, headache secondary to refractive errors, psychogenic headache, dissociative disorder, or anemia.

Table 1: BMI distribution

BMI range (kg/m^2)	Frequency (n=49)
Underweight (<18.5)	0
Normal (<19-24.5)	8 (16.3%)
Overweight (25-29.9)	19 (38.7%)
Obesity class I (30-34.9)	18 (36.7%)
Obesity class II (35-39.9)	2 (4.08%)

Table 2: Clinical Characteristics of our study group

Clinical features	Number of cases (n= 49)
Headache	49(100%)
Transient visual obscuration	23(46.9%)
Double vision	13(26.5%)
Nausea	12(24.5%)
Neck and back pain	12(24.5%)
Vomiting	9(18.4%)
Photophobia	7(14.3%)
Phonophobia	6(12.2%)
Pulsatile tinnitus	5(10.2%)
Dizziness	4(8.2%)
Visual loss	3(6.1)

Systemic diseases-

Twenty four cases (49%) had anemia, oligomenorrhea in 10% of cases, polycystic ovarian syndrome (PCOS) in 11 subjects (22%), and fibroid uterus (2%). Nine patients (18%) have hypothyroidism, diabetes mellitus in 6 (12%) cases and systemic hypertension in 3(6%) patients. Seven cases (10%) were on oral contraceptive pills (OCPs) for their menstrual irregularity.

Neurological examination

Visual acuity was normal in the majority of cases (85.7%). 84 out of 98 eyes had visual acuity of 6/6. Three cases (6/98 eyes) had visual acuity of 6/9 – 6/18, four eyes (4/98) had visual acuity of 6/24-6/60, and 4 out of 98 eyes had a blinding visual loss with visual acuity of <6/60. Pupillary responses were normal in 89.7 %, ill sustained in 10.2%, and there was a relative afferent pupillary defect in one case and lateral rectus palsy in 10

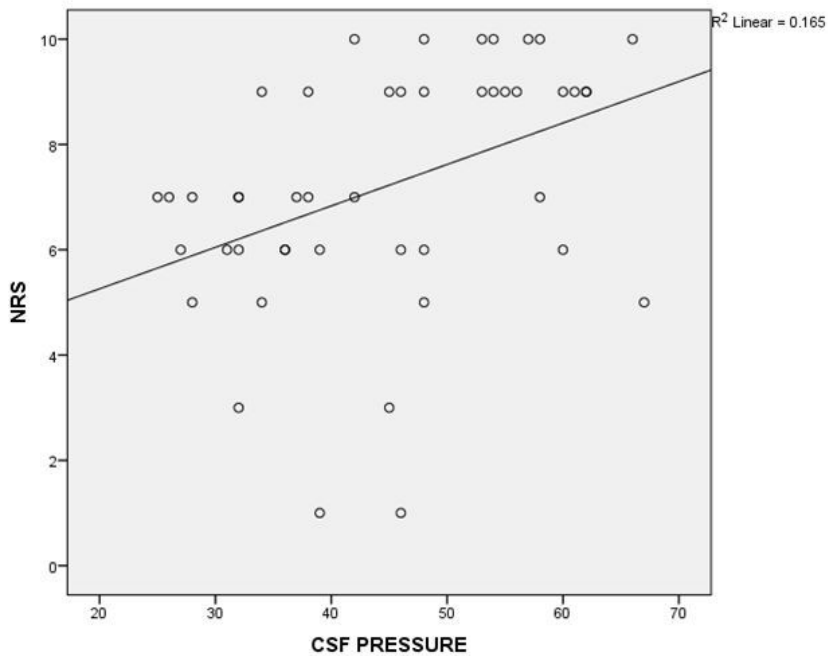

Figure 2. Correlation between CSF opening pressure and headache severity- NRS (Numerical Rating scale)

cases. Visual field defect was noted in 18 cases (36.7%), with enlargement of the blind spot being the most common visual field defect. Other notable defects are nasal, arcuate defects and peripheral field restriction,

Papilledema

A fundus examination was done before starting the therapy. At presentation, bilateral papilledema was noted in 46 (93.8%) cases. Seven cases had modified Frisén grade I papilledema, 8 cases had grade II, 11 cases with grade III, 14 cases had grade IV, and 6 cases had grade V papilledema) and bilateral optic atrophy (secondary) in one (2%) case. Based on the clinical, and radiological findings, as well as high CSF opening pressure remaining three cases were diagnosed as IIH without papilledema (IIHWOP).

The correlation of various clinical parameters with grades of papilledema is summarized in Table 3. Cases of IIH without papilledema (IIHWOP) (grade 0) and low modified Frisén papilledema grades (grades I–III) were grouped. Cases with high modified Frisén papilledema grades (grades IV–V) and secondary optic atrophy were classified together. Abducens nerve palsy, CSF opening pressure, and moderate and severe headache groups showed a statistically significant positive correlation with higher grades of papilledema ($P < 0.05$). The transient blurring of vision and tinnitus did not show a statistically significant difference between the two categories of papilledema mentioned. As illustrated in Figure 3, a higher grade of papilledema showed a positive correlation with higher CSF opening pressure (R^2=0.245, p=0.001). The CSF biochemistry and cytology were within the normal range for all cases. The mean CSF opening pressure was 43.8 ± 12.7 cm H2O. CSF opening pressure was

Table 3: Correlation of various clinical parameters with grades of papilledema

Clinical parameters	Papiledema Grades		Chi square and p value
	Grade I-III (N=26)	Grade IV-V (N=20)	
Moderate headache Numerical rating scale (4-7)			
Yes (n=21)	17 (81%)	4 (19%)	$\chi2$=9.38,df=1
No (n= 25)	9 (36%)	16 (64%)	p =0.002*
Severe headache Numerical rating scale (8-10)			
Yes (n=20)	6 (30%)	14 (70%)	$\chi2$=10.12,df=1
No (n=26)	20 (76.9%)	6 (23.1%)	p=0.001*
Tinnitus			
Yes (n=4)	3 (75%)	1 (25%)	$\chi2$=0.60, df=1
No (n=42)	23 (54.8%)	19 (45.2%)	p=0.435
Transient blurring of vision			
Yes (n=22)	12 (54.5%)	10 (45.5%)	$\chi2$=0.06,df=1
No (n=24)	14 (58.3%)	10 (41.7%)	p=0.796
Abducens Palsy			
Yes (n=12)	4 (33.3%)	8 (66.7%)	$\chi2$=3.55,df=1
No (n=34)	22 (64.7%)	12 (35.3%)	p=0.05*
CSF opening pressure			
<40 mm of H2o (n=19)	16 (84.2%)	3 (15.8%)	$\chi2$=10.09,df=1
≥40 mm of H20 (n=27)	10 (37%)	17 (63%)	p=0.001*

$\chi2$ =chi square, df= degree of freedom, p=p value, n= number of cases
*: Statistically significant

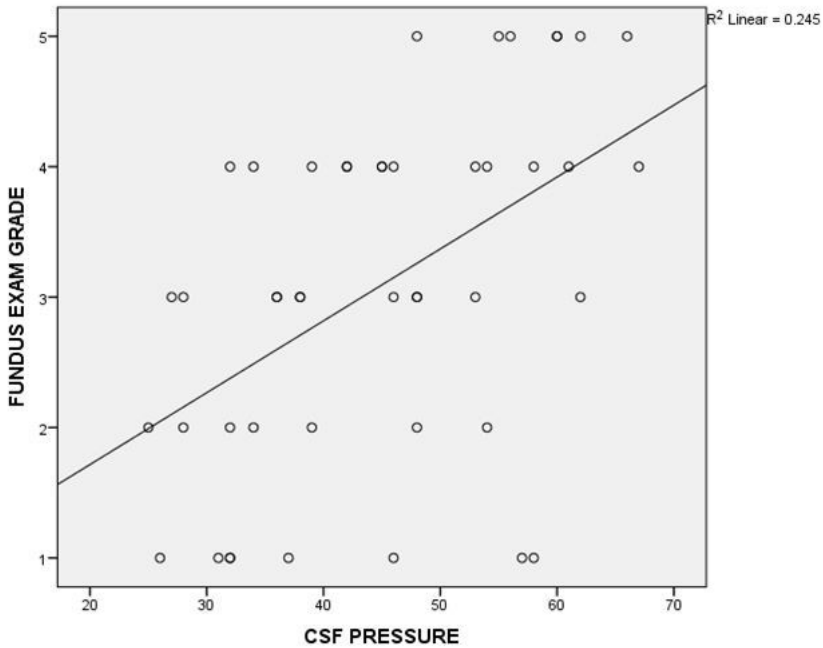

Figure 3. Correlation between CSF opening pressure and Grades of papilledema

recorded <40 cm of H2O in 22 (44.8%) cases and ≥40 cm of H2O was recorded in 27 (55.1%) cases. The correlation of CSF opening pressure values with various clinical features is summarized in Table 4. High CSF opening pressure positively correlated with moderate and severe headache, abducens palsy, and grade IV-V papilledema.

Table 4: Correlation of CSF opening pressure values with various clinical features

Clinical parameters	CSF opening pressure		Chi square and p value
	<40 mm of H_20 (n=22)	≥40 mm of H_20 (n=27)	
Moderate Headache Numerical rating scale (4-7)			
Yes (n= 21)	15 (71.4%)	6 (28.6%)	$\chi2$=10.45,df=1
No (n= 28)	7 (25%)	21 (75%)	p=0.002*
Severe headache Numerical rating scale (8-10)			
Yes (n =20)	2 (10%)	18 (90%)	$\chi2$=16.61,df=1
No (n=29)	20 (69%)	9 (31%)	p= 0.001*
Transient blurring of vision			
Yes (n=23)	12 (52.2%)	11 (47.8%)	$\chi2$=0.92,df=1
No (n= 26)	10 (38.5%)	16 (61.5%)	p=0.338
Tinnitus			
Yes (n=4)	1 (25%)	3 (75%)	$\chi2$=0.69,df=1
No (n=42)	21(46.7%)	24(53.3%)	p=0.404
Abducens palsy			
Yes (n=13)	2 (15.4%)	11 (84.6%)	$\chi2$=6.23,df=1
No (n=36)	20 (55.6%)	16 (44.4%)	p=0.013*
Papiledema grade IV-V			
Yes (n=20)	3 (15%)	17 (85%)	$\chi2$=13.81,df=1
No (n=29)	19 (65.5%)	10 (34.4%)	p=0.001*

$\chi2$ =chi square, df= degree of freedom, p=p value, n= number of cases

*: Statistically significant

Radiological findings

In our series, empty sella was found to be the most common radiological sign (23/49; 46.9%). Other radiological signs were buckling of the optic nerve (17/49; 34.7%), flattening of the globe (20/49; 40.8%), bilateral stenosis of transverse sinuses (16/49; 32.7%), and preoptic space widening (20/49; 41%). Normal MRI brain was found in 7 cases. Empty sella was the only radiological sign which showed a significant correlation with high CSF opening pressure (p- 0.03).

Treatment and follow up

Patients were treated with either medical therapy or medical plus surgical therapy. Acetazolamide was the most common drug used. In two cases Topiramate was used given intolerance to acetazolamide. For four cases acetazolamide was combined with Topiramate given other co-morbidities like morbid obesity and migraine. Three cases underwent ventriculoperitoneal shunt because of rapidly declining visual function. Weight reduction measures were advised for all cases. None of them had undergone bariatric surgery. After treatment patients were followed in the third and sixth months, and findings were summarized in Table 5.

At 6 months follow-up

Two cases were noncompliant with their follow-ups and were excluded.47 cases completed follow-up at the end of 6 months. 60 % of cases had persistent headaches. In the moderate headache group, the severity reduced to 23.6%, in the severe headache group, the severity reduced to 11%. Only 4 cases had persistent visual impairment and visual field defects in 3 cases (6.5%). The papilledema resolved completely in 40 patients (89%), 7 patients had partial improvement of

Table 5: Follow-up of cases at third and sixth month

Clinical Parameters	At diagnosis (n=49)	3 months after treatment (n=49)	6 months after treatment (n=47)
Headache			
Frequency	49 (100%)	38 (77.5%)	28 (60.8%)
Intensity			
Continuous	23 (46.9%)	19 (38.7%)	16 (34%)
Intermittent (>15 days)	20 (40.8%)	15 (30.6%)	9 (32.1%)
Episodic (<15 days)	6 (12.2%)	4 (8.1%)	3 (10.7%)
Site of headache			
Global	11 (42.8%)	9 (18.3%)	7 (14.9%)
Bitemporal	4 (8%)	2 (4%)	1 (2.1%)
Unilateral	12(22.4%)	10 (20.4)	9 (19.1%)
Occipital	9 (18.4%)	7 (12.2%)	3 (6.4%)
Occipitofrontal	13 (26.5%)	10 (20.4%)	8 (17%)
Severity of headache-VAS			
Mild (1-3)	7 (14.2%)	11 (22.4%)	8 (17%)
Moderate (4-7)	22 (44.9%)	12 (24.4%)	9 (19.1%)
Severe (8-10)	20 (40.9%)	15 (30.6%)	11 (23.4%)
No headache	0	10 (20.4%)	18 (38.2%)
Visual impairment	10 (20.4%)	5 (10.2%)	4 (8.5%)
Visual field defects	18 (36.7%)	6 (12.2%)	3 (6.5%)
Papilledema grade 0	3 (6.1%)	26 (54.1%)	40 (85.1%)
Papilledema grade 1	7 (14.2%)	5 (10.4%)	3 (6.3%)
Papilledema grade 2	8 (16.3%)	11 (22.9%)	2 (4.2%)
Papilledema grade 3	10 (20.4%)	3 (6.25%)	0
Papilledema grade 4	14 (28.6%)	2 (4.1%)	0
Papilledema grade 5 and optic atrophy	7 (12.2%)	2 (4.1%)	2 (4.2%)

their papilledema grade, one patient had residual high grades of papilledema (GV), and only one had optic atrophy. In patients who underwent ventriculoperitoneal shunting; one out of three had optic atrophy at the end of 6 months of follow-up. Other patients had partial improvement in visual function. There was no significant correlation (p-value = 0.21) between the follow-up grade of papilledema and the patient's BMI. No significant correlation between the amount of weight lost and the improvement in VAS score at 6 months (p value = 0.32).

DISCUSSION

In our study population majority were females (98%). Most large IIH trials report a female preponderance of around 90%.[8-10] This high prevalence suggests that IIH is more common in women and that most males have other illnesses such as sleep apnea-related intracranial hypertension. The most common age group of onset was from 16 to 62 years, with a mean age of 29 ± 9.7 years, which is consistent with the results of previously published papers.[8-10] The relationship between obesity and IIH has been well described in the literature.

The risk of IIH in women increases due to obesity, as it affects their hormonal state by increasing adipose tissue mass. However, the role of hormonal differences in men and women with IIH is not yet fully established. We found, 38 % were overweight and 44% were obese and 16 % had a normal BMI. In our study, the WHO definition of BMI was used to maintain consistency of definition across the studies.

A BMI standard specifically for Asian Indians has been proposed to address the alarming prevalence of diabetes and cardiovascular disease within this community. It takes into account the various correlations between BMI and body fat.[11] Accordingly, the patients were categorized as underweight (<18.5 kg/m2), normal or lean BMI (18.5–22.9 kg/m2), overweight (23.0–24.9 kg/m2), and obese (≥25 kg/m2). When this criterion of BMI was applied, 82% of patients in our cohort fell into the overweight/obese category; this aligns with the research conducted by Gafoor and colleagues.[12] Therefore, the ethnic-specific BMI cut points should be implemented for all obesity-related health issues. Although many Western literatures reported a significant proportion of obesity among IIH cases, our study and a few other Indian studies found a significant number of cases with normal BMI.[12,13] Sugarman *et al*.[14] suggested that central obesity increases intra-abdominal pressure, leading to increased cardiac filling pressure and impeded venous return from the brain, which in turn results in increased intracranial venous pressure and raised ICP. Akin to the previous studies, our study also showed a statistically insignificant relationship between opening pressure and BMI.[12,15] The current study showed PCOS in 37% which is comparable with analysis by Glueck *et al*. who reported a prevalence rate of 39% in IIH cases.[16]

Headache was the most common initial symptom (100%), which is consistent with other studies. Headache was constant and daily in 46.9% which is similar to that described in earlier series.[8,17-19]

At the end of 6 months, our study demonstrated that headaches have been significantly reduced in 40% of cases. The headache was relieved after 3 months in 23% of cases. No history of headache was present in more than half of the patients who reported daily persistent headaches at 6 months of follow-up, which is most likely a de novo chronic headache caused by IIH. Persistent headaches after normalization of intracranial pressure have been reported even more frequently.[17,19] Ekizoglu *et al*. proposed that IIH may trigger sensitization of central pain pathways, similar to primary headaches.[20] The raised CSF pressure might lead to unrelenting trigeminal activation that persists even if the intracranial pressure is normalized. The chronic dysregulation of normal mechanisms that suppress inappropriate trigeminal nociceptive signals could occur as a result of trigeminal dysfunction, which is similar to migraine headaches.[21] Our results were in line with the conclusion that headaches can persist despite papilledema resolution, supporting the hypothesis that raised ICP is not the sole mechanism of headache in IIH.[18] Headache is often multifactorial, which can be either due to increased intracranial pressure, medication overuse, migraine, or a combination of these. Hence we hypothesize that headache is a poor marker of disease activity. The headache phenotype in IIH is heterogeneous with subjects having headaches similar to migraine, tension type headache. Albeit there are no specific features for headache in IIH, associated symptoms like transient visual obscuration, diplopia, and pulsatile tinnitus aid in diagnosing IIH rather than a primary headache disorder. However, in 18% of the individuals headache is the sole presenting feature and none of these associated symptoms were present. This accentuates the importance of fundus examination while evaluating a case of headache, especially in obese childbearing females.

Multiple investigations that included patients with IIH found that transient visual obscurations were the second most frequent symptom, which was also true of our sample. In agreement with previous studies[22,23] our study also found that blurred vision and diplopia were the predominant visual symptoms. IIH is a major contributor to vision impairment and is no longer regarded as benign. Antecedent studies have shown that a small but considerable number of individuals with IIH may be at risk for developing irreversible visual impairment.[22,23] In the early 1980s, Wall *et al*. reported a visual loss of 77.5% using automated perimetry. Corbett et al followed up 57 patients for 5 to 41 years and found that 24% of them developed severe visual impairment.[22] However several recent studies (western as well as Indian) have exhibited good visual prognosis in the long term[9,19,24,25], which is identical to our study. With the advent of sophisticated neuroimaging techniques, early diagnosis, and treatment could be a reason for less visual impairment in these cohorts. In the current study, only 4 cases (8%) were left with severe visual impairment. The temporal profile of visual impairment in IIH is capricious. Most develop gradually with deficits ranging from a few weeks to months, while at other times vision impairment can occur very early. Hence long-term visual follow-up is warranted in IIH cases.

We found a significant positive correlation between the severity of headaches and CSF opening pressure. This trend is similar to Sultan *et al*, a group of investigators who followed IIH patients for 3 months in an Egyptian cohort. However, this is contrary to Friedman's study in the idiopathic intracranial treatment trial (IIHTT), where 165 newly diagnosed cases were followed for 6 months, and they found no correlation between headache severity and CSF opening pressure.[26] This lack of association between the severity of headaches and CSF opening pressure may be due to natural fluctuation in the CSF pressure throughout the day.[27]

Bilateral papilledema was found in the majority of the cases (96 %), which is analogous to previous studies.[8-10] At the end of 6 months, papilledema was completely resolved in 85% of cases. In agreement with the former series[10,24], papilledema grade was significantly correlated with CSF opening pressure. Several Indian studies also have shown similar findings.[28-30]

Empty sella was the most common radiological finding, which is consistent with earlier studies.[24,30] However, few studies[13,28] reported flattening of posterior sclera as the most common finding. It has been well documented in the literature that none of the MRI brain and MRV findings is specific for IIH Interestingly, when CSF pressure has been normalized and papilledema has resolved, most neuroimaging findings do not improve.[19] We did not find any correlation between radiological features and CSF opening pressure which is comparable to the study done by Sharma *et al*.[30] None of the patients had any recurrence or worsening of symptoms on a follow-up which has been found in some studies[8,12], the probable reason could be the short duration of follow-up in our cohort. In the current study, only 3 cases required ventriculoperitoneal shunting. The majority of the cases responded well to medical management alone. The strength of this study is that there are few long-term studies in Asia, and most Indian studies are retrospective.

We acknowledge the limitations of our study, the sample size was small and the period of follow-up is up to 6 months. Follow-up does not include OCT monitoring and other parameters. Data were compared from baseline to 6 months follow-up, which may not provide the actual picture, as IIH is a chronic disorder and relapses/recurrences are well known.

In conclusion, headache is the most prominent feature of IIH. Patients with IIH can have coexisting primary headaches. Differentiation of IIH from primary headache is essential as management differs completely. Persistence of headache even after the resolution of papilledema suggests that headache in IIH is multifactorial. Further studies are warranted in this regard because headache is a major disability in IIH. The overall outcome for vision is good. We found there was a significant positive correlation between CSF opening pressure, the severity of a headache, and the grade of papilledema. IIH should be suspected in any obese woman with headaches since it is one of the preventable causes of blindness. Long-term follow-up of IIH cases is mandatory as they are prone to recurrence.

DISCLOSURE

Financial support: None

Conflict of interest: None

REFERENCES

1. Durcan FJ, Corbett JJ, Wall M. The incidence of pseudotumor cerebri: population studies in Iowa and Louisiana. *Arch Neurol* 1988;45(8):875-7. DOI: 10.1001/archneur.1988.00520320065016

2. Wakerley BR, Tan MH, Ting EY. Idiopathic intracranial hypertension. *Cephalalgia* 2015;35(3):248-61. DOI: 10.1177/0333102414534329
3. Shah VA, Kardon RH, Lee AG, Corbett JJ, Wall M. Long-term follow-up of idiopathic intracranial hypertension: the Iowa experience. *Neurology* 2008;70:634-40. DOI: 10.4103/0972-2327.78044
4. Rowe FJ, Sarkies NJ. Assessment of visual function in idiopathic intracranial hypertension: a prospective study. *Eye* 1988;12:111. DOI: 10.1038/eye.1998.18
5. Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. Neurology. 2002;59(10):1492-5. DOI: 10.1212/01.wnl.0000029570.69134.1b.
6. https://www.who.int/europe/news-room/fact-sheets/item/a-healthy-lifestyle---who-recommendations.
7. Jensen MP, Karoly P, O'Riordan EF, *et al*. The subjective experience of acute pain. An assessment of the utility of 10 indices. *Clinical J Pain* 1989; 5:153-9. DOI: 10.1097/00002508-198906000-00005
8. Wall M, Kupersmith MJ, Kieburtz KD, *et al*. The idiopathic intracranial hypertension treatment trial: clinical profile at baseline. *JAMA Neurol* 2014;71(6):693-701. DOI: 10.1001/jamaneurol.2014.133.
9. Ambika S, Arjundas D, Noronha V, Anshuman. Clinical profile, evaluation, management and visual outcome of idiopathic intracranial hypertension in a neuro-ophthalmology clinic of a tertiary referral ophthalmic center in India. *Ann Indian Acad Neurol* 2010;13:37-41. DOI: 10.4103/0972-2327.61275
10. Kattah JC, Pula JH, Mejico LJ, McDermott MP, Kupersmith MJ, Wall M. CSF pressure, papilledema grade, and response to acetazolamide in the idiopathic intracranial hypertension treatment trial. *J Neurol* 2015;262:2271-4. DOI: 10.1007/s00415-015-7838-9.
11. WHO Expert Consultation. Appropriate body-mass index for Asian populations and its implications for policy and intervention strategies. *Lancet* 2004;363:157-63. DOI: 10.1016/S0140-6736(03)15268-3
12. Gafoor VA, Smita B, Jose J. Long-term response of cerebrospinal fluid pressure in patients with idiopathic intracranial hypertension – A prospective observational study. *Ann Indian Acad Neurol* 2017;20:220-4. DOI: 10.4103/aian.AIAN_32_17
13. Pal A, Sengupta P, Biswas D, Sen C, Mukherjee A, Pal S. Pattern of idiopathic intracranial hypertension in Indian population. *Ann Indian Acad Neurol* 2019;22:47-51. DOI: 10.4103/aian.AIAN_160_19.
14. Sugarman HJ, DeMaria EJ, Felton WL 3rd, Nakatsuka M, Sismanis A. Increased intra-abdominal pressure and cardiac filling pressures in obesity-associated pseudotumor cerebri. *Neurology* 1997;49:507-11. DOI: 10.1212/wnl.49.2.507
15. Corbett JJ, Mehta MP. Cerebrospinal fluid pressure in normal obese subjects and patients with pseudotumor cerebri. *Neurology* 1983;33:1386-8. DOI: 10.1212/wnl.33.10.1386
16. Glueck CJ, Iyengar S, Goldenberg N, Smith LS, Wang P. Idiopathic intracranial hypertension: associations with coagulation disorders and polycystic-ovary syndrome. *J Lab Clin Med* 2003;142(1):35-45. DOI: 10.1016/S0022-2143(03)00069-6.
17. Yri HM, Wegener M, Sander B, Jensen R. Idiopathic intracranial hypertension is not benign: a long-term outcome study. *J Neurol* 2012;259(5):886-94. DOI: 10.1007/s00415-011-6273-9.
18. Yri HM, Rönnbäck C, Wegener M, Hamann S, Jensen RH. The course of headache in idiopathic intracranial hypertension: a 12-month prospective follow-up study. *Eur J Neurol* 2014;21(12):1458-64. DOI: 10.1111/ene.12512.
19. Xu W, Prime Z, Papchenko T, Danesh-Meyer HV. Long term outcomes of idiopathic intracranial hypertension: Observational study and literature review. *Clin Neurol Neurosurg* 2021;205:106463. DOI: 10.1016/j.clineuro.2020.106463.
20. Ekizoglu E, Baykan B, Orhan EK, Ertas M. The analysis of allodynia in patients with idiopathic intracranial hypertension. *Cephalalgia* 2012; 32(14):1049-58. DOI: 10.1177/0333102412457091.
21. Boyer N, Dallel R, Artola A, Monconduit L. General trigeminospinal central sensitization and impaired descending pain inhibitory controls contribute to migraine progression. *Pain* 2014;155: 1196-205. DOI: 10.1016/j.pain.2014.03.001
22. Wall M, George D. Visual loss in pseudotumor cerebri: incidence and defects related to visual field strategy. *Arch Neurol* 1987;44:170-5. DOI: 10.1001/archneur.1987.00520140040015
23. Corbett JJ, Savino PJ, Thompson HS, *et al*. Visual loss in pseudotumor cerebri. Follow-up of 57 patients from five to 41 years and a profile of 14 patients with permanent severe visual loss. *Arch Neurol* 1982;39:461-74. DOI: 10.1001/archneur.1982.00510200003001
24. Sultan LI, Elnekidy AM, Elfatatry AM, *et al*. A clinical and radiological study in patients with idiopathic intracranial hypertension. *Egypt J Neurol Psychiatry Neurosurg* 2020;56:71. https://doi.org/10.1186/s41983-020-00189-w.
25. Baheti NN, Nair M, Thomas SV. Long-term visual outcome in idiopathic intracranial hypertension. *Ann Indian Acad Neurol* 2011;14(1):19-22. DOI: 10.4103/0972-2327.78044.
26. Friedman DI, Quiros PA, Subramanian PS, *et al*. Headache in idiopathic intracranial hypertension: findings from the idiopathic intracranial hypertension treatment trial. *Headache* 2017;57(8):1195-205. DOI: 10.1111/head.13153.
27. Johnston I, Paterson A. Benign intracranial hypertension. II. CSF pressure and circulation. *Brain* 1974;97(2):301-12. DOI: 10.1093/brain/97.1.301
28. Byju N, Mathew DS, Biju PR. A study on the clinical profile of Idiopathic Intracranial Hypertension (IIH) in a tertiary care centre in India. *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)* 2021;20(11):33-40.
29. Dubey A, Athale S. A clinical profile of Idiopathic Intracranial Hypertension (IIH) in a tertiary referral teaching centre in Central India. *Indian J Neurosci* 2017;3(1):36-40. DOI: 10.18231/2455-8451.2017.0006
30. Sharma B, Seervi N, Sharma V, Panagariya A, Goel D. Clinical and radiological profile of 122 cases of idiopathic intracranial hypertension in a tertiary care centre of India: An observational study. *Neurol India* 2022;70(2):704. DOI: 10.4103/0028-3886.344644