ORIGINAL RESEARCH

Ophthalmological manifestations and clinical features of idiopathic intracranial hypertension in adults at a tertiary care center in Rajasthan, India: A cross-sectional study

[1]Dr. Shilpi Kochar, [2]Dr. Abhishek Kochar

[1]Associate Professor, Department of Ophthalmology, S.P.Medical College, Bikaner, Rajasthan, India
[2]Medical Officer, Department of Neurology, S.P.Medical College, Bikaner, Rajasthan, India

Corresponding Author
Dr. Shilpi Kochar
Associate Professor, Department of Ophthalmology, S.P.Medical College, Bikaner, Rajasthan, India

Received: 22 March, 2022 Accepted: 26 April, 2022

ABSTRACT

Purpose: The study aimed at describing the ophthalmological manifestations of patients with idiopathic intracranial hypertension (IIH) and corelate severity of papilledema with cerebrospinal fluid (CSF) opening pressure. **Methods:** This was a prospective cross-sectional study. Patients aged 20-60 years using modified Dandy criteria diagnosed with IIH were included. Demographic details, ocular symptoms and signs were noted and papilledema graded. Investigations included Visual field analysis and CSF opening pressures were noted. **Results:** The study included 20 patients of average age 35.64 (± 7.87) years with a predominantly female population (95%). Mean BMI was 34.76 kg/m^2(±5.32) kg/m^2. Common presenting complaint was headache (87.5%). The most common biochemical abnormality was Vitamin D deficiency (45%). The visual status of most patients was 6/6 BCVA on Snellen's Chart (60%). Lateral rectus palsy was present in 15%. Papilledema was present in 85 % eyes with 30% showing Grade I papilledema. There was visual field loss in 52.5% of study eyes. Mean CSF opening pressure was 292.51(±114.47) mmH_2O. MRI showed empty sella (35%) and tortuous optic nerve (20%) empty sella (34.4%). MRV elucidated most frequent abnormality as transverse sinus stenosis. No correlation was seen between CSF opening pressure and grade of papilledema and severity of VF defect or BMI. **Conclusion:** Clinical profile of patients with IIH was a female with high BMI of reproductive age with headache. Visual field examinations are important tool for planning follow-up.

INTRODUCTION

Idiopathic intracranial hypertension (IIH) as the name suggests is of unknown etiology, affecting mostly women with high BMI of childbearing age. Intracranial pressure (ICP) is elevated with neurological manifestation of papilledema, leading to optic atrophy.[1]

The diagnosis of IIH is based on the modified Dandy criteria.[2] The estimated annual incidence is 0.9 per 100,000 in the general population with a female-to-male ratio of 8:1.[3]

Our aim was to describe the clinical features of IIH, eye manifestations and correlate CSF opening pressure with grading and severity to help in elucidating features that can lead to early diagnosis, treatment and improvement of visual prognosis.

METHODS

A prospective, cross-sectional, descriptive study was carried out between October 2019and March2020. All consecutive patients diagnosed with IIH as per the modified Dandy criteria between ages 20 and 60 years were included. Patients with any other coexisting eye pathology, neurological symptoms and ones who were incapable to undergo Visual fields were excluded.

The demographic data included age and sex, a comprehensive history including the onset and chronicity of symptoms, past medical history, history of any drugs or medications, personal and family history. Symptoms of headache, tinnitus, dimunition of vision, double vision, light and sound sensitivity were recorded.

Body mass index (BMI) was calculated after accurate height and weight measurements. Detailed eye examination included visual acuity with Snellen's chart, near vision with Times New Roman chart and color vision with Ishihara's pseudo-isochromatic test plates. Anterior segment examination with a slit-lamp biomicroscope, pupillary reactions and extraocular motility were assessed. Fundoscopy was done with a direct ophthalmoscope, an indirect ophthalmoscope and +78 D slit-lamp biomicroscopy. Papilledema was graded according to the Frisen grading system.[3]

Visual fields (30-2 Threshold Swedish Interactive Threshold Algorithm (SITA) Standard) were performed using Humphrey field analyzer. Visual fields defects, mean deviation and visual field index were recorded. Visual field defects were graded according to Wall and George criteria,[4] which included grade I consisting of enlarged blind spot with inferior nasal step; grade II, enlarged blind spot with peripheral nasal loss; grade III, arcuate defects; grade IV, double-arcuate defects arcing around central 10 degrees; and grade V, gradual depression of the visual field more pronounced peripherally.

Opening CSF pressure measured on lumbar puncture in lateral decubitus position was recorded with detailed CSF analysis findings.

Magnetic resonance imaging (MRI) and/or magnetic resonance venogram (MRV) were done to rule out any CNS pathology and to look for optic nerve. Blood investigations included complete blood count, Vitamin B 12 levels, Serum Iron and folic acid and Vitamin D levels to look for any association with IIH

Sample size was calculated using nMaster: Proportion in the population taken as 0.9 and sample population of 0.7 with power of the study 80% and alpha error 5%; sample size was calculated as 18.

RESULTS

A total of 20 patients diagnosed with IIH were analysed during the study period fulfilling the inclusion criteria. The age range was 20–60 years with a mean(±SD) of 35.64 (± 7.87) years. Females accounted for a majority of patients (19/20, 95%). BMI was in the range of 18.13–42.60 kg/m^2 (mean(±SD) of 34.76 kg/m^2 (±5.32) kg/m^2).

Headache was seen in 17/20 (85%) whiledimunition or blurring of vision in 9/20 (45%) patients and these were the two most frequent complaints. Other symptoms were transient visual obscurations (TVO) (35%), double vision (35%), intolerance to light (25%), tinnitus (15%), nausea (15%), intolerance to sound(10%) and CSF rhinorrhea (5%).

In 2/20 patients (10%), there was history of long-term intake of medications from non allopathic sources. There was positive history of oral contraceptive pills (OCPs) in 10% and other hormones in 10%. Vitamin D deficiency was seen in blood investigations in 9/20 patients (45%). Low Hemoglobin levels were seen in 8/20 patients. Other comorbidities seen were low vitamin B12 levels (35%) and hypothyroidism (20%).

A total of 24/40 (60%) eyes of total patients in the study had 6/6 visual acuity. Color vision was normal in 36/40 eyes (90%). Normal Pupillary reactionwere seen in 85% of eyes, ill-sustained/RAPD was seen in 15%, and double vision was demonstrated in 7/20 patients while lateral rectus palsy was seen in 3 (15%) of them.

Many patients (20/40) had signs of long standing papilledema on fundus examination. Papilledema was absent in 15% of eyes, and there was optic atrophy in 4 eyes out of 40 (10%). In the remaining eyes (90%) grading of papilledema was done as per Frisen criteria.

Visual field assessment showed a mean deviation in the right eye of the range − 9.84 to +0.29 dB (mean(±SD) of −2.89(±2.48) dB) and in the left eye of the range of −24.38 to +0.2 dB (mean(±SD) of −4.88(±4.96) dB) [Table 1]. 19 of 40 eyeseyes (47.5%) had no visual field loss and field loss severity was grade I in 20%, grade II in 10%, grade III in 12.5 %, grade IV in 5% and grade V in 5% of eyes. No statistically significant correlation was found between the grade of papilledema and mean deviation of visual fields.

Lumbar puncture was done in 19/20 patients with a CSF opening pressure range of 120–910 mmH_2O (mean(±SD) of 292.51(±114.47) mmH_2O.

The most common findings on MRI were empty sella (7/20 ; 35%), dilated optic sheath or prominent perioptic CSF spaces (2/20, 10%), tortuous optic nerve (4/20, 20%) and posterior flattening of sclera (2/20, 10%). MRI was normal in 40% of patients. Of the 11 MRV (Venogram) done, transverse sinus stenosis was the most frequent finding.

DISCUSSION

The mean age in our study was35.64 years with total study population between 20-60 years. In the idiopathic intracranial hypertension treatment trial (IIHTT),[5] mean age of the study population was 29 years. Studies by Ambika et al.,[2] Ayush Dubey et al.[6] and Claire Chagot et al.[7] had a study population with a mean age of 32 years, 30 years and 33 years, respectively.

Gender distribution was majorly skewed with female predilection occupying 95% of study population. This was similar to the IIHTT trial (97.6% females),[5] and and Claire Chagot et al.[7] (92.4% females). Ayush Dubey et al.[6] had all female patients in their study while the female population was only at around 80% in Studies by Ambika et al.[2]

A majority patients in our study were overweight with a mean BMI of 34.76 kg/m^2, as compared to 35 kg/m^2 in the Claire Chagot et al.[7] study and mean BMI was 39.9 kg/m^{2}in IIHTT,[5].

Family history of IIH in our study was not positive for any patient, while in the IIHTT,[5] it was 5%. The above mentioned demographics concerning IIH from research studies show IIH is a disease of adult females

with high BMI who are of reproductive age group with no family history.

The most frequent complaint at presentation was headache (85%) followed by dimunitionof vision (45%). Other symptoms were transient visual obscurations (35%), diplopia (35%), nausea (15%), photophobia (25%), tinnitus (15%) and phonophobia (10%). One patient presented with CSF rhinorrhea. Lateral Rectus Palsy was seen in 3 patients out of 7 patients who had diplopia.The complaints of intolerance to sound and CSF rhinorrhea were not reported in the other studies.

The most common presentation in the IIHT trial,[5] Ambika et al.,[2] Ayush Dubey et al.[6] and Claire Chagot et al.[7] studies wa also Headache with the incidence of headache being 84%, 94%, 92.9%, 82.3% respectively. An incidence of transient visual obscurations (68%), pulse synchronous tinnitus (52%), non-pulsatile tinnitus (23%) which is higher than our study was documented in The IIHTT study.[5] whereas the incidence of diplopia (18%) was low. In the study by Ambika et al.,[3] occurrence of TVO and tinnitus was 68% and 58%, respectively. This was higher when compared to our study population. The other symptoms were sustained visual loss (26%), flashes (54%), double vision (38%) and pain behind the eyeball (44%).[3]

The study by Ayush Dubey et al.[6] reported blurring of vision (78.6%), diplopia (57.1%) and tinnitus (50%). 42.8% of cases were reported to suffer from lateral rectus palsy. Claire Chagot et al.[7] reported TVO (17.7%), eye tracking impairment (15.2%), tinnitus (12.7%) and dizziness (11.4%). Hence, comparing the data the incidence of transient visual obscurations and diplopia was lower while occurrence of tinnitus was similar to our study.[7]

A case report of IIH in a patient with megaloblastic anemia was presented by T Van Gelder et al.[8] the etiology of association is still not understood but low Hemoglobin could be a predisposition.. Vitamin D deficiency has not been reported in anyprevious study. As with other studies, our study also noticed history of alternative medicine usage, OCPs and Hormone replacement therapies present in such patients. Reports in the past have shown that history of corticosteroid withdrawal and hormone replacement therapy can precipitate IIH.[1] Other common comorbidities found were vitamin D deficiency (45%), anemia (40%), vitamin B12 deficiency (35%) and hypothyroidism (20%). The mean hemoglobin level was found to be 10.9 g/dl with mean vitamin B12 levels at 339.33 pg/ml and mean vitamin D levels at 13.10 ng/dl.

Our study found that most eyes had a visual acuity of 6/6 (60%) which was similar to that found in the IIHTT (70.9% in study eyes and 77.0% in fellow eyes)[5] and Ambika et al.[2] (55%). Ambika et al.[3] also reported 6/9–6/18 visual acuity in 20%, 6/24–6/60 in 7% and <6/60 in 18%. 30% eyes had visual acuity between 6/9 and 6/18, 5% eyes between 6/24 and 6/60 and 5% less than 6/60 in our study.

No correlation was found between BCVA with the degree of visual field loss in our study. Visual field loss was seen in 52.5% of eyes. The IIHTT[5] reported that the most common perimetric finding was a partial arcuate scotoma with enlarged blind spot. However, the inclusion criterion was that the study eye should have a mild degree of visual field loss in IIHTT.[5] Therefore, their perimetric results might not be representative of visual loss of IIH in general. Normal visual field was noted in 12% of cases, nasal and arcuate defects in 7% and advanced generalized constricted fields in 11%in Ambika et al.[2]. Similar to studies in the past , our study also reported eyes with BCVA as 6/6 with a visual field loss, thus postulating visual field rather than visual acuity as a fairermarker and functional test of visual loss in IIH.

Another clinical sign assessed in our study was pupillary reaction. It was brisk in 85%, ill-sustained/ RAPD in 15%. The IIHTT[5] showed RAPD in 5.4% eyes. It was postulated that optic atrophy points to chronicity of disc edema and occurs due to misdiagnosis and delay in treatment.

Our study eyes presented with papilledema in most cases (85%): grade I in 30 %, grade II in 20%, grade III in 20 %, grade IV in 5% and grade V in 5%. There was no papilledema in 15%, and 10% had progressed to optic atrophy. Grade I papilledema was the most frequent presentation in our study in contrast to the IIHTT,[5] which had grade II as the most common presentation.Likewise, grade II papilledema was most common (28.6%) reported by Ayush Dubey et al.[6].The grade of papilledema and grade of visual field defect Showed did not show any statistically significant correlation.

In our study, the mean CSF opening pressure was 292.51(±114.47) mmH_2O. This was lower than the IIHTT,[5] where the mean was 343.5 mmH_2O. In the study by Claire Chagot et al.,[7] median CSF opening pressure was 285 mmH_2O (range 150–540) mmH_2O which was closer to our study. CSF opening pressure is essential for the diagnosis of IIH. In our study, there was no statistically significant correlation between CSF opening pressure and perimetry mean deviation. No correlation was also found between BMI and CSF opening pressure as well.

MRI findings in our study were normal in 40%. MRI findings were found to be associated with IIH in medical literature,[7] and the findings present in our study population were empty sella in 35% of cases, dilated optic sheath in 10%, tortuous optic nerve in 20%, posterior flattening of sclera in 10% and all of the above in 5%. In the study by Ambika et al.,[3] MRI was done in 42/50 patients out of which 25% were normal, perioptic space widening in 14 and empty sella in 3 patients. Ayush Dubey et al.[6] reported normal MRI scans in 85.7%, empty sella in 7.1% and distended optic nerve sheath in 7.1%.

MRV was performed in 11 patients and wasabsolutely normal in 53%. Transverse sinus stenosis was seen in 10 % of patients in our study and the reports were not similar to the studies by Ambika et al.[2] and Ayush Dubey et al.[6] in which all were normal. Abnormal neuroimaging was the exclusion criteria in Ayush Dubey et al.[6] Unilateral transverse sinus stenosis or hypoplasia in 17% and bilateral transverse sinus stenosis (bilateral stenosis or unilateral stenosis and hypoplasia) in 74% of cases was reported by Chagot et al.,[7] They postulated transverse sinus stenosis plays a role in pathogenesis of IIH.

CONCLUSION

Idiopathic Intracranial Hypertension if not diagnosed, investigated and treated meticulously can potentially be vision threatening. Our study results show that IIH is a disease in overweight females of reproductive age group with the most frequent presenting complaint as headache. The study has also shown that anyone with high BMI on any suspicious drugs or hormone therapy are also predisposed and hence should be thoroughly examined and investigated if bilateral disc edema present and diagnosis of IIH should be kept in mind. Perimetry is a fair indicator of the the optic nerve functionality and prognosticationas it detects visual field loss before fall in visual acuity. It can serve as an important tool for early diagnosis, intervention and management. Papilledema grading and its chronicity are potential indicators of prognosis of visual status and planning follow-up.

Ethical clearance

Approval was obtained from the Institutional Ethics Committee.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Daroff RB Bradley's Neurology in Clinical Practice 7th ed Philadelphia Elsevier Saunders 2016
2. Ambika S, Arjundas D, Veena 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
3. Miller NR, Newman NJ, Blousse V, Kerrison JB Walsh And Hoyt's Clinical Neuro-Ophthalmology 6th ed United States of America Lippincott Williams and Wilkins 2005
4. Wall M The importance of visual field testing in idiopathic intracranial hypertension Continuum (MinneapMinn) 2014 20 1067 74
5. Wall M, Kupersmith MJ, Kieburtz KD, Corbett JJ, Feldon SE, Friedman DI, et al. The idiopathic intracranial hypertension treatment trial:Clinical profile at baseline JAMA Neurol 2014 71 693 701
6. Dubey A, Athale S A clinical profile of Idiopathic Intracranial Hypertension (IIH) in a tertiary referral teaching centre in Central India IP Indian J Neurosci 2017 3 36 40
7. Chagot C, Blonski M, Machu JL, Bracard S, Lacour JC, Richard S Idiopathic intracranial hypertension:Prognostic factors and multidisciplinary management J Obes 2017 2017 5348928
8. Van Gelder T, van Gemert HM, Tjiong HL A patient with megaloblastic anaemia and idiopathic intracranial hypertension:Case history Clin Neurol Neurosurg 1991 93 321 2
9. Passi N, Degnan AJ, Levy LM MR imaging of papilledema and visual pathways:Effects of increased intracranial pressure and pathophysiologic mechanisms AJNR Am J Neuroradiol 2013 34 919 24