



A Case Series on Clinical Profile, Risk Factors and Management of Idiopathic Intracranial Hypertension

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Introduction

Idiopathic intracranial hypertension (IIH) occurs when there is raised intracranial pressure (ICP) of unknown etiology and is diagnosed when all other causes of raised ICP have been excluded¹. IIH most commonly affects women of child-bearing age and overweight women². However, men, women of all ages and children of both genders are affected³. Previously the terms like pseudo tumor cerebri and benign intracranial hypertension were used; however, in view of impending loss of vision it cannot be considered benign. Hence the term IIH is used nowadays. The diagnosis of IIH was made using modified Dandy criteria which includes presence of raised intracranial pressure, papilloedema, normal magnetic resonance imaging (MRI) and elevated CSF opening pressure with normal CSF composition. In the absence of papilledema, the diagnosis is challenging and requires specific neuroimaging criteria to be fulfilled.

Objectives

To study the risk factors and management of IIH.

Materials

This is a case series of nine patients who got admitted in neuromedicine ward between January 2021 and June 2022 with symptoms and signs suggestive of idiopathic intracranial hypertension. A detailed history and clinical examination including detailed fundus examination were done. Every patient underwent magnetic resonance imaging (MRI) of brain and magnetic resonance venogram, CSF manometry, and routine CSF analysis. Diagnosis of IIH was made using the modified Dandy criteria.

Details on age at onset of symptoms, sex, height, weight, body mass index (BMI), presence of comorbid conditions, use of medications, findings of ophthalmic examination including fundoscopy, neurological features, CSF opening pressure, radiological signs on cerebral MRI (prominent subarachnoid space around the optic nerves, flattening of the posterior sclera, partial empty sella, and slit-like ventricles, presence of transverse sinus stenosis on MRV) and type of medical treatment were recorded.

All patients were treated medically with oral acetazolamide at a dose of 0.5-1 g/day and comorbidities were managed accordingly.

Case Reports

Case 1

A 27-year-old female presented on 10/11/21 with a 2 months history of severe headache and blurring of vision for 1 month. Two weeks prior to presentation she started experiencing headaches associated with tinnitus and TVOs and her symptoms were worse in the morning. No history of vomiting. On examination her Body Mass Index (BMI) was 31.3. Her blood pressure was 136/90mmHg. Her visual acuity was 6/9 in both eyes. The fundus examination revealed established papilloedema. MRI brain report was suggestive of IIH. CSF opening pressure was 410 mm of H₂O and 25 ml of CSF was drained during lumbar puncture. CSF biochemistry and cytology analysis came to be normal. Subsequently she was found to have hypothyroidism. Her abdominal ultrasound imaging was normal. She was commenced on oral acetazolamide 250mg 3 times daily, L-thyroxine and advised weight reduction. Patient improved symptomatically on treatment.

Case 2

A 35-year-old female presented on 12/10/21 with history of headache of 3 months duration which was associated with blurring of vision and vomiting. Associated with TVO and tiredness. On examination her Body Mass Index (BMI) was 27.3. Her blood pressure was 130/80mmHg. Her visual acuity was 6/12 in both eyes. Patient was found to have microcytic hypochromic anemia and her haemoglobin was 6.8 g/dL. The fundus examination revealed chronic papilledema. MRI brain report was suggestive of IIH with empty sella and hypoplastic left transverse sinus and sigmoid sinus. CSF opening pressure was 290 mm of H₂O and 20 ml of CSF was drained during lumbar puncture and CSF analysis was normal. Abdominal ultrasound and thyroid function test were normal. She was commenced on oral

acetazolamide 250mg 3 times daily and iron supplementation.

Case 3

A 36 year-old female got admitted with 1 month history of headache and with double vision for 2 weeks. She had associated transient visual obscurations (TVOs) and tinnitus. She was a diagnosed case of hypothyroidism on irregular treatment. On examination her BMI was 28.3. Her blood pressure was 120/70mmHg. The visual acuity was 6/6 in both eyes and colour vision was normal. Examination of the optic discs revealed established papilledema. Her haemoglobin was 8.1g/dL and peripheral smear showed microcytic hypochromic picture. A lumbar puncture was done and the opening pressure was found to be 310 mm of H₂O. Results of her CSF biochemistry and cytology were normal. MRI brain was suggestive of IIH. She was treated with acetazolamide, iron and thyroxine supplementation.

Case 4

A 23 year-old female presented with headache of 15 days duration which was holocranial and was associated with early morning head ache. The patient had a history of irregular menses for which she was on oral contraceptive pills (OCP). On examination her blood pressure was 110/80 mm of Hg. Her BMI was normal. The visual acuity was 6/9 in right eye and 6/6 in left eye and colour vision was normal. Her fundus examination showed bilateral established papilledema. Lumbar puncture was performed and the opening pressure was found to be 330 mm of H₂O with normal CSF analysis. 25 ml of CSF was drained during procedure. MRI brain was suggestive of IIH. Thyroid function test and ultrasound scanning of abdomen were normal. She was treated with acetazolamide and improved symptomatically.

Case 5

A 40 year old female got admitted in neurology department with complaints of headache of 1 month duration and associated vomiting. Patient had transient visual obscurations as well. On examination her BP was 120/70 mm of Hg and

had a BMI of 29.8. Her visual acuity was found to be 6/6 in both eyes. Her fundoscopy revealed bilateral established papilloedema. Thyroid function test was normal. MRI brain showed prominence of perioptic CSF space, bulging of optic disc and narrowing of left transvers sinus. CSF opening pressure was 280 mm of H₂O and 30 ml of CSF was drained during lumbar puncture. Patient was started on oral acetazolamide with other symptomatic treatment.

Case 6

A 25 year old male presented with history of headache, blurring of vision and transient visual obscuration of 6 weeks duration. Associated with early morning headache. On examination his blood pressure and BMI were within normal range. He had a visual acuity of 6/12 and 6/9 on right and left eyes respectively. Fundus examination showed bilateral established papilledema. His MRI brain was suggestive of IIH with an elevated opening pressure. During lumbar puncture 20 ml of CSF was removed and CSF analysis was normal. Patient was initiated on acetazolamide 250 mg TID. The patient's symptoms improved on treatment.

Case 7

A 47 year old female got admitted with headache of 1 month duration, blurring of vision and double vision of 2 weeks duration and tiredness. This was associated with early morning headache. She had past history of migraine. On examination her BP was 120/76 mm of Hg, and BMI was 28.4. Her visual acuity was 6/12 on both eyes and fundus showed bilateral papilledema. Her haemoglobin was 7.8g/dL and peripheral smear was showing microcytic hypochromic anemia. MRI brain was suggestive of IIH. CSF opening pressure was 290 mm of H₂O, and 25 ml of CSF was removed. Biochemical cytological analysis of CSF was normal. She was treated with acetazolamide and iron supplementation, following which she improved symptomatically.

Case 8

A 39 year old female who is a known case of migraine presented with headache of 5 months duration, which became holocranial with associated transient visual obscuration. She also complained of early morning headache for last 2 months. On examination her BP was 130/76 mm of Hg, visual acuity was 6/9 and 6/6 on right and left eyes respectively. Her fundus examination did not show any evidence of papilledema. Her haemoglobin was 5.9 with a microcytic hypochromic picture. Thyroid function test and ultrasound abdomen were normal. MRI brain was suggestive of IIH and was confirmed with CSF opening pressure which was 280 mm of H₂O. During lumbar puncture 30 ml of CSF was drained. Subsequently patient was started on iron supplementation and acetazolamide. Patient was discharged with symptomatic improvement.

Case 9

A 45 year old female was admitted with persistent diffuse headache with associated vomiting and double vision of one month duration. On examination her BP was 130/84 mm of Hg and BMI of 24. Visual acuity was 6/6 in both eyes. Bilateral abduction restriction was present. Fundus examination showed bilateral established papilledema. MRI brain was suggestive of IIH. Her total WBC count was 87200, platelet count of 6.3 lakhs and haemoglobin of 12.7 g/dL. Liver and renal function tests were within normal limits. Ultrasound abdomen showed mild hepatosplenomegaly. CSF opening pressure was 400 mm of H₂O with normal biochemical and cytology reports. 25 ml of CSF was let out during lumbar puncture. Peripheral blood smear showed feature of chronic myeloid leukemia in chronic phase with Philadelphia chromosome positivity. Patient was treated with mannitol, acetazolamide, imatinib and allopurinol.

Results

Out of the nine cases reported to our institute, eight were females and age between 23-47 yrs. All

presented with complaints of headache with a duration of 1 month to 5 months. Headache was associated with transient visual obscuration in 5 cases, diplopia in 2 cases, blurring of vision in 5 cases. Eight patients had bilateral papilledema. Best-corrected visual acuity range from 6/12 to 6/6. CSF opening pressure was in the range of 280 -410 mm of H₂O. Four patients had microcytic hypochromic anemia, 2 patients had hypothyroidism. Five patients had BMI >25 and one patient was on oral contraceptive pills. In all case MRI brain was suggestive of IIH and MRV had ruled out cortical venous thrombosis. Patients were treated with Acetazolamide at a dose of 0.5 – 1 g/day. Four patients with microcytic hypochromic anemia were treated with oral iron supplementation, two patients with hypothyroidism received levothyroxine and one patient with CML received chemotherapy apart from acetazolamide.

Variables	Mean
Gender	
Male	1
Female	8
Age	35.2 yrs
BMI	
<18.5	0
18.5 - 25	4
25-30	4
>30	1
Symptoms and signs at presentation	
Headache	9
TOVs	5
Tinnitus	1
Blurring of vision	5
Diplopia	3
Papilledema	8
Risk Factors	
BMI >25	5
Anemia	4
Obstructive sleep apnea	1
OCP	1
Vitamin A	0
Hypothyroidism	2
CML	1
Comorbidities	
Systemic Hypertension	0
Diabetes mellitus	0
Migraine	2
Others	0

Discussion

Several studies have shown that IIH is more common in women and obese individuals⁴. Among women aged 20–44 years who are 20% or more above ideal body weight, the incidence increases to about 20 times the incidence in the general population⁴⁻⁵. In this series out of the nine case eight were females (89%). Various previous reports has shown more frequency of female cases as 89%- 97.57%⁶. In this series, age group of presentation was 23 to 47 years with mean age of 35.2 years. . In many previous reports, mean age of presentation ranged between 29 to 32 years.⁶ Obesity has a strong association with IIH in most cases and weight loss alleviates IIH signs and symptoms significantly while weight gain is linked to recurrence⁷. It has been observed that even a modest degree of weight loss (5-10%) is usually required for improvement in symptoms and signs^{8,9}. In our series five patients had overweight.

Symptoms in our series were similar to those found in other previous studies^{10,11}. Headache was the most common initial symptom in our patients (100%). Transient visual obscuration (TVO) was seen in 5 patients (56%). Other studies had reported TVO in 60.4% -72%^{10,12}. Even though tinnitus was reported in 50–60% of cases in previous studies^{13,14}, in our series only one patient had tinnitus (11%). Two case of our series had diplopia (22%). Diplopia was reported in 18 -30% of cases with IIH in previous studies^{15,16}

Bilateral papilledema was found in all of our patients except for one who had IIH without papilledema and was diagnosed based on elevated opening pressure and neuroimaging. Digre et al. reported IIH without papilledema in 5.7% of cases¹⁷.

Patients with IIH frequently have coexisting obstructive sleep apnea (OSA). Previous studies suggest that upto 60% of IIH patients have OSA¹⁸. In our series only one patient (11.1%) had OSA.

Two patients of this series had history of migraine (22%). The prevalence of migraine in patients with IIH is about 31.8%¹⁹

One patient gave the history of oral contraceptive pill intake. Khin .P et al in their study had stated that hormonal contraceptives were not significantly associated with a higher incidence of IIH²⁰.

In our series four patients had microcytic hypochromic anemia (44.4%). Pooled incidence of anemia in patients with IIH was found to be 18.2% in a meta analysis²¹. Another patient was later diagnosed as a case of chronic myeloid leukemia. Raised WBC count can lead to hyperviscosity of blood which in turn interfere with CSF absorption in subarachnoid granulations.

In this series none of the patients had systemic hypertension or diabetes mellitus. Previous studies had shown a 25 % prevalence of systemic hypertension or diabetes mellitus in patients with IIH⁶.

All patients were treated medically with oral acetazolamide at a dose of 0.5-1 g/day. A Cochrane systematic review identified two randomised control trials for the use of acetazolamide in IIH²². Topiramate has been reported to be efficacious in relieving headache caused by IIH.²³

Conclusion

Apart from usual risk factors for IIH, clinicians should search for less common risk factors as well and provide appropriate treatment to prevent the risk of permanent visual loss

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