

Case Report

Idiopathic Intracranial Hypertension Presenting with Multiple Cranial Nerve Palsy

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Idiopathic Intracranial Hypertension (IIH) or Pseudotumor cerebri is an uncommon neurological disease of unknown aetiology. It is characterized by elevated intracranial pressure without apparent aetiology and associated with normal Cerebrospinal fluid analysis without any structural lesion of the brain. Patients usually present with Headache, transient blurring of vision and occasional involvement of Cranial Nerve, most commonly 6th Cranial Nerve. But involvement of Multiple Cranial Nerves in Idiopathic Intracranial Hypertension is rare. We are presenting a case of a 20 years old female admitted with history of Headache, Blurring of vision and Diplopia, and was diagnosed as pseudotumor cerebri with Multiple Cranial Nerve Palsy (left sided 6th and right sided incomplete 3rd Cranial Nerve Palsy). Our patient improved on treatment with acetazolamide. Physician should be aware of Pseudotumor cerebri. Early diagnosis and treatment may improve patients' vision.

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Key words : Idiopathic Intracranial Hypertension, Multiple Cranial Nerve Palsy, Rare Association.

Pseudotumor cerebri or Idiopathic Intracranial Hypertension (IIH) is an uncommon Neurological Disorder of unknown aetiology. It is characterized by features of elevated Intra Cranial Pressure (ICP) and high opening pressure of Cerebrospinal fluid without structural lesions of the brain¹. Primarily it occurs in women with higher Body Mass Index (BMI), Obstructive Sleep Apnoea or Autoimmune Aetiology². The incidence rate in the general population is 1.56 cases / 100,000 / year and 11.9 / 100,000 / year in overweight and obese women³. The pathophysiology of IIH is not clear. Overweight or Obesity, Hypertension, Autoimmune Diseases, Uremia, Over Dosages of Drugs (eg, Vitamin A, Tetracycline, Minocycline, Nalidixic Acid, or Oral Contraceptive Pills), and chronic respiratory insufficiency are common associated risk factors of IIH. Most common presentation is headache (92%) and two-third of patients complains of transient blurring of vision⁴. 6th Cranial Nerve (CN) is commonly affected that was reported in 12% of adults and 9-48% of children. But involvement of multiple CNs in IIH is very rare⁵. We are presenting a case of IIH with multiple CN palsies (left sided 6th and right sided incomplete 3rd Cranial Nerve Palsy).

CASE REPORT

A 20 years old female presented to the Emergency Department of Midnapore Medical College on 24th December, 2021 complaining of severe holocranial headache for 7 days. Her headache was insidious onset and gradually progressive and was associated with

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Editor's Comment :

- IIH should be suspected in overweight and obese patients presenting with headache and transient blurring of vision.
- IIHs usually presents with headache, blurring of vision and bilateral 6th cranial nerve palsy but involvement of multiple cranial nerves are not uncommon.
- Fundoscopy should be routinely done in all patients presenting with sudden onset headache, as visual acuity may be normal initially.
- Early diagnosis and timely commencement of treatment can save the vision of patient.

Vomiting, Weakness, Dizziness and Transient Blurring Of Vision. On 4th day of her illness, horizontal diplopia with maximal separation of images on left direction of gaze had developed. She denied any history of Fever, Convulsion, Weakness of any Limb, Trauma or Tick Bite. She was normotensive and non-diabetic. There was no significant past medical history or family history of similar illness. On general physical examination patient was conscious, alert and cooperative, BMI was - 35.2 kg/m², Blood Pressure - 130/80 mm of Hg, Pulse Rate - 74/min, Respiratory Rate - 18 / min without pallor, cyanosis, clubbing, oedema and icterus. She was not able to abduct her left eye (Fig 1) and there was limitation of extorsion of her right eye (Fig 2). Pupillary reaction was normal with a visual acuity of 6/6 in both the eyes and normal colour vision. Fundoscopy showed bilateral Papilledema (Fig 3). Rest of the neurological examination, including sensory and motor function, reflexes, coordination and gait was normal. The systemic examination was unremarkable.

Hemogram, Routine and Microscopic examination of Urine, Electrolytes, Liver Function Tests, Urea and Creatinine, T3, fT4 and TSH, autoimmune profile and D-dimer were within normal limits. Computed Tomography



Fig 1 — Loss of abduction of left eye consistent with left 6th cranial nerve palsy



Fig 2 — Loss of extorsion movement of right eye as a secondary action of inferior rectus due to right sided

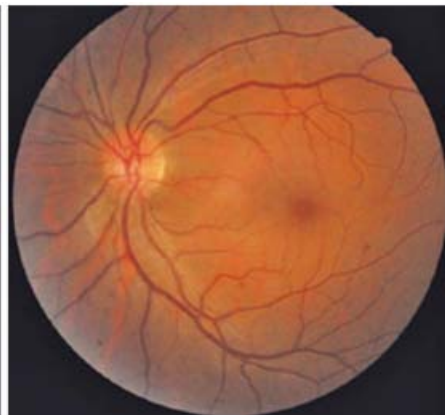


Fig 3 — Fundoscopy showing bilateral optic disc swellings

(non-contrast) was normal. Cerebrospinal fluid study reported normal except high opening pressure (265 cm of H₂O). Magnetic Resonance Imaging (MRI) of brain (Fig 4) and Magnetic Resonance Venogram were normal.

IIH is diagnosed by exclusion of other causes and neuro-imaging studies should be performed to rule out the structural lesions of the brain. Normal neuro-imaging of brain excluded the structural and vascular lesions in our case. Use of drugs like Tetracycline, high dose of Vitamin A, corticosteroid, lithium and Oral Contraceptive Pills were excluded in our patient. Only involvement of multiple CNs without other manifestation of Sarcoidosis and normal Leptomeninges on MRI make the diagnosis of Neurosarcoidosis unlikely. In absence of history of Tick Bite, Fever and abnormal CSF analysis, the diagnosis of Lyme Disease in our case is less likely.

Normal neuro-imaging supported the diagnosis of IIH as the most probable cause of the patient's symptoms. The diagnosis was further supported by the marked improvement with acetazolamide therapy.

The patient was treated with oral Acetazolamide 500 mg twice daily and Paracetamol and discharged with the same medication. On follow-up after 1 month, the patient was symptomatically better without headache and visual problems (Table 1), and there was improvement of ocular movements (Fig 4) with partial resolution of papilledema.

DISCUSSION

IIH is an uncommon Neurological Diseases with signs and symptoms of elevated ICP without structural lesions of the brain. In 1897, Quincke first described IIH⁶. IIH was named Pseudotumor cerebri in 1904⁷. Headache and Transient Blurring of vision are the commonest presentation of

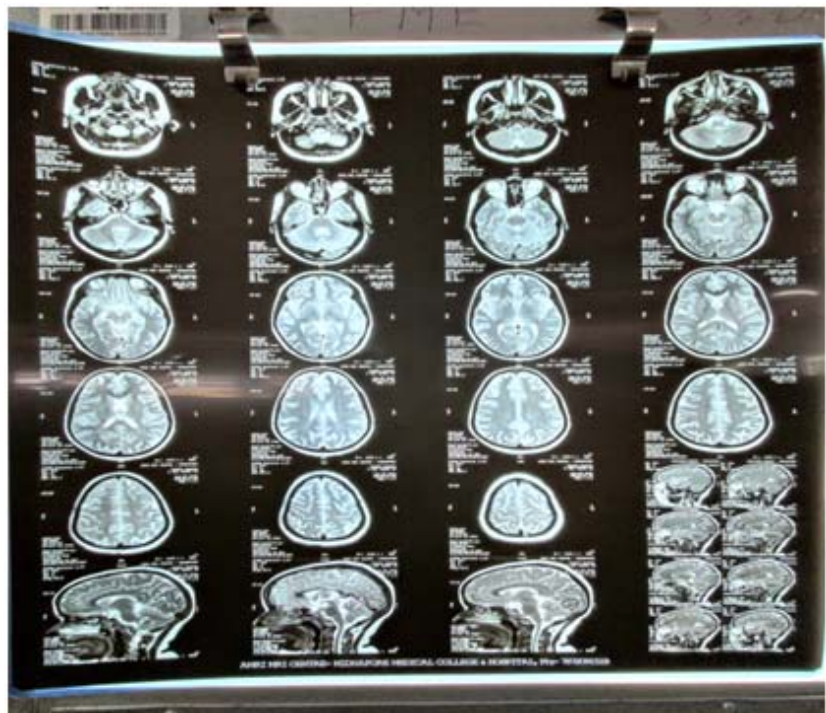


Fig 4 — MRI of brain showing normal sized ventricles



Fig 5 — Improved ocular movements on follow up visit

IIH^{8,9}. Headache is the most frequent symptom. Females are most commonly affected. More than 90% patients with IIH are either overweight or obese and in child bearing age group. IIH is diagnosed by the presence of all the criteria (Friedman's criteria) {normal neurological examination except involvement of CN (usually 6th CN), Papilledema, elevated Lumbar Puncture (LP) pressure ≥ 250 cm H₂O and normal CSF study without any structural abnormality of the brain}. It is often associated with non-localizing neurologic symptoms, other than unilateral or bilateral 6th CN palsy. Multiple CNs involvement in IIH was rarely reported in the literature. Our patient of IIH presented with left sided 6th and right sided incomplete 3rd Cranial Nerve Palsy. Modified Dandy criterion for IIH do not allow for any focal neurological deficit other than 6th Nerve Palsy but our case report showing Multiple Cranial Nerve Palsy indicating the need to revise it.

The pathophysiology of involvement of multiple CNs in IIH is not exactly known. It is a false localizing sign due to elevated ICP^{10,11}. Due to its long intracranial course, 6th Cranial Nerve is usually susceptible to elevated ICP. Raised ICP, along with venous congestion of the brain parenchyma or Ocular Motor Nerves of 3rd and 4th CN, had been suggested as possible aetiologies¹². But the possibility of coincidental association could not be completely refuted in our patient.

Agarwal, *et al* reported 3rd, 6th and 7th CN involvement in IIH who improved with dexamethasone and LP¹³. Soroken C, *et al* reported a case of IIH with multiple CNs palsies in a 13 years old girl who completely recovered within 3 weeks of treatment with acetazolamide¹⁴. A 16 years old girl presented with bilateral 6th, and left 3rd and 4th CN Palsies secondary to IIH in primary antiphospholipid syndrome¹⁵.

The major complication of IIH is papilledema that may result in loss of vision. Acetazolamide decreases CSF production by inhibiting carbonic anhydrase and commonly prescribed to treat IIH.

In conclusion, IIH is an uncommon neurological

Day	Headache	Visual Blurring	Diplopia	3 rd and 6 th Cranial Nerve Palsy	Papilledma
Day 1	+	+	-	+	+
Day 4	+	+	+	+	+
Day 7	-	+	+	+	+
Day 14	-	+	+	+	+
Day 30	-	-	-	+/-	-

disorder. Loss of vision is main long term complication of IIH. Early diagnosis and treatment may improve patients' vision.

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