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A Race against Time: Fulminant Idiopathic Intracranial Hypertension and Optic Atrophy

[1]Dr. Nishika Jain, [2]Dr. Aneesha Trivedi*, [3]Dr. Saurav Patil, [4]Dr. Akarshan Mehta, [5]Dr. Harsha Pagad, [6]Dr. Saroj Sahdev

[1,2,3,4]Third Year Junior Resident, [5]Assistant Professor, [6]Professor & HOD,
Department of Ophthalmology,
D.Y. Patil School of Medicine & Hospital, Nerul, Navi Mumbai

***Corresponding Author:**
Dr. Aneesha Trivedi
Third Year Junior Resident, Department of Ophthalmology,
D.Y. Patil School of Medicine & Hospital, Nerul, Navi Mumbai.

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Abstract

This article describes the case of an18-year-old woman with fulminant IIH. Fulminant IIH is a dreaded condition with rapid deterioration of vision happening in few days to few weeks .The article concludes that early intervention with acetazolamide and surgical procedures such as shunting, optic nerve sheath fenestration, or ventricular shunting increase the chance of successful visual recovery in individuals with fulminant IIH. There are still a subset of patients who may not respond to the conventional treatment with irreversible loss of vision. Our patient belonged to the same with bilateral optic atrophy and blindness.

Keywords: Fulminant idiopathic intracranial hypertension, papilloedema, optic atrophy

Introduction

Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri, is a neurological disorder characterized by elevated pressure within the brain, in the absence of any structural abnormalities. It is most commonly seen in obese women of reproductive age, with peak incidence occurring in the second to fourth decades of life. However, in a small percentage of individuals with IIH, rapidly increasing vision loss can occur within a month of the onset of symptoms, leading to a risk of permanent blindness. The main symptom of IIH is papilloedema, which is bilateral and symmetrical swelling of the optic nerve head. The aim of treatment for IIH is to identify and address the underlying cause of the headache, through a combination of medical and surgical therapies.

Case Report:

An 18 year old female came to a major tertiary care center in Navi Mumbai with chief complaints of severe headache and diminished vision in both eyes since six months. She had no documented systemic illness.

On general examination, the patient is well nourished and weighed 55kgs. Her right eye unaided visual acuity was hand movement close to face and left eye was finger counting at one meter.

Colour vision could not be assessed.

On pupillary evaluation, there was sluggish response to light in both eyes. Right eye revealed a grade 2 RAPD. Anterior segment examination was unremarkable. Intraocular pressure by applanation tonometry was 12 mmHg in both eyes.

On Hirschberg reflex test, 15 degree exotropia was seen in the right eye. Ocular motility was normal.

Posterior segment evaluation on slit lamp biomicroscope using a 90D lens and indirect ophthalmoscopy revealed bilateral optic disc pallor

with sheathing of vessels with cuff of peripapillary fluid.

On examination of previous medical records, it was found that patient was diagnosed with papilloedema (right eye more than left) 4 months back. The MRI brain done at the time, reported partially empty sella and prominence of the infratentorial CSF spaces, cerebellar foliae suggestive of mild cerebellar atrophy. In view of above findings patient was subjected to lumbar puncture.

CSF analysis was normal. The opening pressure was more than 25 cm water. Patient was started on oral acetazolamide 250 mg thrice a day. In view of persistent symptoms and worsening vision, a lumboperitoneal shunt was performed.

On OCT ONH & RNFL, average RNFL thickness was 49 µm in both eyes.

On OCT macula ganglion cell complex analysis, average and minimum GCL + IPL thickness were significantly reduced.

Given the possibility of peripapillary fluid, a USG B scan was ordered which showed bilateral minimal enlargement of the optic nerve posterior to the optic disc. (possibility of fluid in the meninges) Patient was referred to neurologist for evaluating the patency of shunt and further management.

Figure 1. Right and left eye fundus on indirect ophthalmoscope showing optic disc pallor

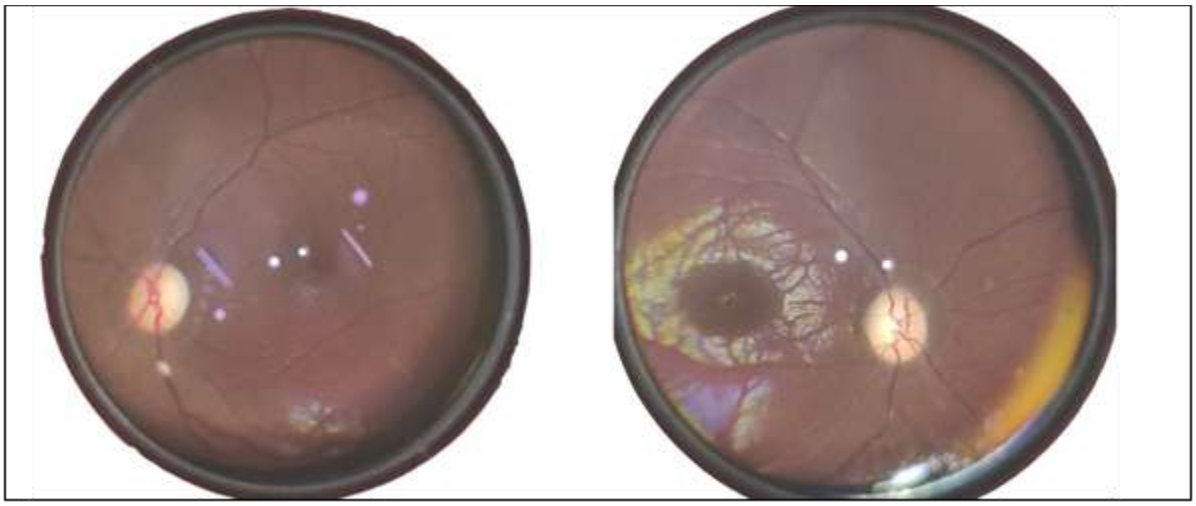

Figure 2: MRI Brain

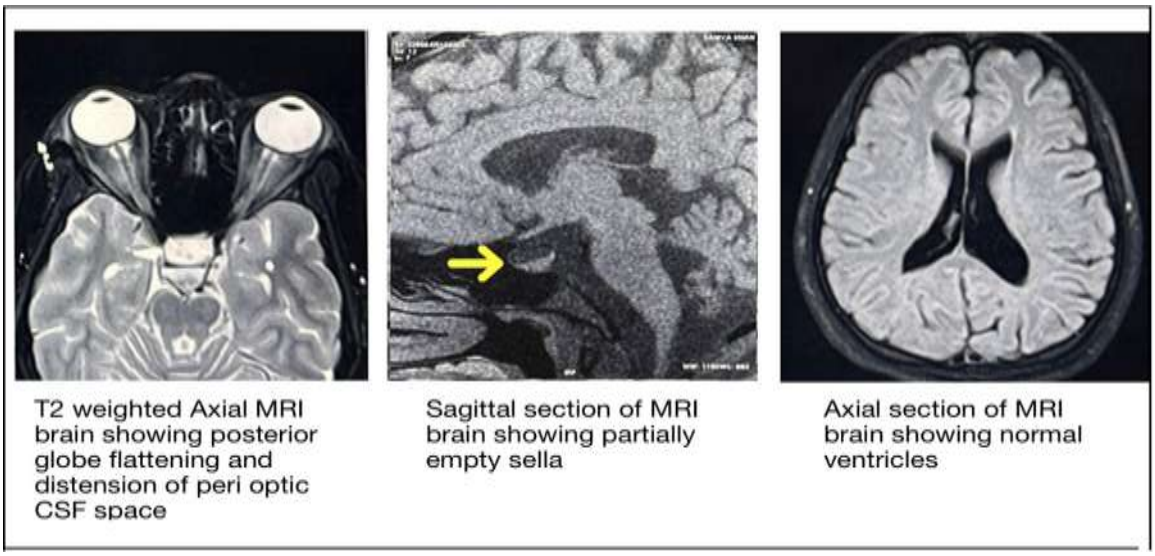

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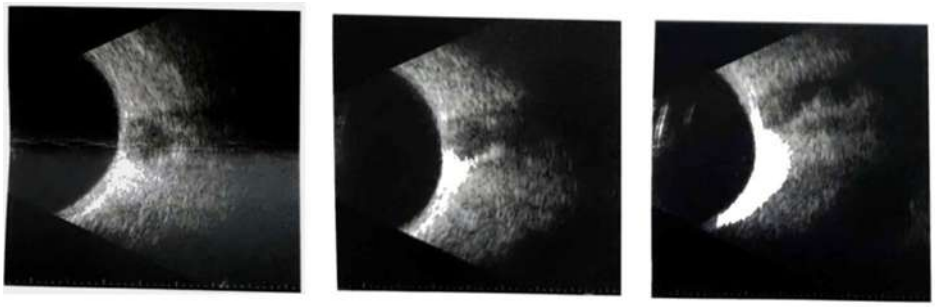
Figure 3: USG B scan suggests possibility of fluid in the meninges



Discussion

Fulminant idiopathic intracranial hypertension (IIH) is a rare but serious medical condition characterized by a sudden and severe increase in intracranial pressure. It affects 2-3% of IIH patients and can cause rapidly progressing visual loss within one month of the beginning of IIH symptoms. It is more likely to affect overweight or obese individuals(1), pregnant women, and those taking hormonal birth control or certain medications like tetracycline antibiotics. The cause of IIH is not well understood, but it is believed to be related to an imbalance between the production and absorption of cerebrospinal fluid (CSF) in the brain.

Fulminant IIH can cause severe headache, pulsatile tinnitus, and temporary vision loss.(2-6) A diagnosis of IIH is made through a combination of medical history, physical examination, imaging tests, and a measurement of intracranial pressure. Bedside ocular ultrasonography (USG B scan) is a non-invasive and portable technique that can quickly measure the optic nerve sheath diameter (ONSD) and identify elevated ICP. (8) USG B scan is also useful in detecting an increased size of the optic nerve head and in the assessment of subarachnoid fluid around the optic nerve. (9) ONSD has also been used to monitor the treatment efficacy in patients with IIH.(10)

The diagnostic criteria for fulminant IIH include fulfilling the criteria for IIH with papilledema, less than 4 weeks between symptom onset and severe loss of visual acuity or field, and rapid worsening of vision over days. (2) (7) Fulminant IIH can also occur in atypical populations for IIH, including men and children.

Two case reports, by Bhandohal and by Shaikh et al., describe patients who had normal vision initially, but rapidly developed blindness in one or both eyes within a short period of time. These cases highlight the importance of seeking immediate medical attention if there is a sudden change in symptoms due to fulminant IIH.(7)

Treatment for IIH may include weight loss, medications to reduce fluid production, and a lumbar puncture to remove excess CSF. In some cases, surgery may be necessary to relieve pressure on the optic nerve and prevent vision loss. However, the management of fulminant IIH differs fundamentally from the management of typical IIH. Patients suspected to have fulminant IIH should receive rapid diagnosis confirmation and medical and surgical management, as delaying intervention could lead to permanent visual loss. Early involvement of a neuro-ophthalmologist is crucial, and patients with progressive visual loss should be closely monitored until effective treatments are found.

Effective treatment of fulminant IIH requires a combination of medical and surgical interventions, including high dose carbonic anhydrase inhibitors and intravenous methylprednisolone as temporary measures, as well as CSF shunting, optic nerve sheath fenestration (ONSF), or venous sinus stenting as surgical interventions. If surgical intervention is delayed or rapid vision loss has occurred, temporizing CSF drainage should be considered through lumbar or extraventricular drains or serial large volume lumbar punctures.

Our case presented with a severe headache and was diagnosed with benign IIH. She received oral acetazolamide and underwent a lumbar peritoneal shunt, but remained symptomatic. The patient was later diagnosed with optic atrophy secondary to fulminant IIH. Early intervention is important to increase the chances of visual recovery.

Acetazolamide, at a dose of 2-4g/day, should be initiated while awaiting surgical intervention, which may include shunting, ONSF, or ventriculoperitoneal shunt (VSS). In our case, early ONSF could have

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salvaged the patient's vision and prevented the progression of optic atrophy.

Conclusion:

In conclusion, fulminant IIH is a rare but potentially vision-threatening condition, and prompt intervention is crucial to prevent permanent visual disability. Further studies are needed to better understand the underlying mechanisms and to improve the management of this condition.

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