Resolution of Fulminant Idiopathic Intracranial Hypertension Treated with Acetazolamide

Ojas Srivastava, Jonathan A. Micieli

MD Program, University of Alberta, Edmonton, AB, Canada; Department of Ophthalmology and Vision Sciences, University of Toronto, Toronto, ON, Canada; Kensington Vision and Research Centre, Toronto, ON, Canada; Department of Ophthalmology, St. Michael’s Hospital, Unity Health, Toronto, ON, Canada

Keywords

Idiopathic intracranial hypertension · Fulminant intracranial hypertension · Hemorrhagic optic disk edema

Abstract

Idiopathic intracranial hypertension (IIH) is a condition of elevated intracranial pressure commonly seen in obese women of childbearing age. Fulminant IIH is a rare subset of IIH that is characterized by rapidly progressive vision loss in less than 4 weeks, and typically requires surgical intervention for treatment. We describe a 36-year-old man with a 3-week history of acute onset vision loss and fulminant IIH in whom severe bilateral hemorrhagic optic disk edema was identified. There were also associated moderate visual field defects. Given the rapid onset of symptoms and severity of papilledema, surgical management was discussed but the patient had opted for medical management and close follow-up. He began oral acetazolamide, which was escalated to the maximal dose of 4 g and seen regularly with close follow-up. Four months after presentation, he was completely symptom free and the bilateral optic disk edema had resolved. His visual fields had also improved. We emphasize the importance of close follow-up in fulminant IIH and highlight that although most cases often require surgical intervention, some patients may show improvement with medical management only.
Introduction

Idiopathic intracranial hypertension (IIH) is a syndrome characterized by elevated intracranial pressure of unknown etiology and normal cerebrospinal fluid (CSF) contents seen most commonly in obese women of childbearing age [1]. Fulminant IIH is characterized by having IIH with an acute onset of symptoms less than 4 weeks between symptom onset and vision loss and worsening of visual function over this period of time [2]. Given the rapid onset of symptoms and imminent risk of permanent vision loss, fulminant IIH is considered an urgent situation that requires immediate management. Surgical options such as CSF shunting, optic nerve sheath fenestration, or venous sinus stenting are often required given the serious nature of the visual deficits. Herein, we present a case of fulminant IIH with hemorrhaging of the optic disks in a man who recovered significantly with only medical treatment with acetazolamide.

Case Report

A 36-year-old man was referred for bilateral vision loss. He had a past medical history of obesity (BMI 47.3), obstructive sleep apnea, and depression. He took sertraline and was compliant with CPAP. He denied use of vitamin A derivatives or tetracyclines. He reported a 3-week history of new onset blurred vision in both eyes with a new constant holocephalic headache. He denied pulsatile tinnitus, diplopia, or recent change in weight. He initially presented to an optometrist and was referred urgently to neuro-ophthalmology after the discovery of severe, hemorrhagic optic disk edema. Initial neuro-ophthalmology exam revealed a visual acuity of 20/25 OD and 20/30 OS, and Humphrey 24-2 SITA-Fast visual fields showed a superior arcuate defect in the right eye (MD −18.88 dB) and an enlarged blind spot and nasal step in the left eye (MD −11.57 dB, Fig. 1a). Color vision was 12/14 OD and 11/14 OS Ishihara color plates. Dilated fundus exam showed severe, hemorrhagic optic disk edema (Fig. 2a). He was thought to have severe papilledema, and he was admitted to hospital for a CT/CTV of the head that showed signs consistent with raised intracranial pressure including bilateral distal transverse sinus stenosis. Lumbar puncture in left lateral decubitus position revealed

Fig. 1. Humphrey 24-2 SITA-Fast visual fields at presentation (a) and final follow-up (b) 6 months later. The mean deviation at presentation was −18.88 dB OD MD −11.57 dB OS and at final follow-up was −2.21 dB OD and −3.19 dB OS.
an opening pressure of 45 cm of water with normal CSF contents. A total of 15 cc was removed, and the closing pressure was not measured. Complete blood count, creatinine, and electrolytes were normal. Given the severe amount of vision loss, surgical intervention such as transverse sinus stenting of CSF shunt was discussed, but the patient wanted to think about it and opted for short interval follow-up and medical therapy. He started on oral acetazolamide 1 g twice daily and was seen in follow-up 1 week later. At that follow-up, he felt subjectively improved and the visual acuity, visual field, and optic disk edema were mildly improved. He declined surgical intervention, and acetazolamide was increased to 2 g twice daily. He continued at this dose, and at the 1-month follow-up, his visual field improved and the mean deviation was now −8.79 dB OD and −7.62 dB OS. He continued on this high dose for a total of 3 months, and at that time, his visual acuity was 20/25 OD and 20/25 OS. Humphrey 24-2 SITA-Fast visual field testing showed several depressed points in both eyes with a mean deviation of −5.79 dB OD and −4.87 dB OS, the optic disk edema was resolved, and there was mild optic disk pallor (Fig. 2b). Acetazolamide was discontinued at that time. His final follow-up was at 6 months after presentation, and at that time, his visual acuity was 20/20 OD and 20/20 OS. Humphrey 24-2 SITA-Fast visual fields were performed and are shown in Figure 1b. The mean deviation was −2.21 dB OD and −3.19 dB OS with a foveal threshold of 37 dB OD and 37 dB OS. The optic nerve appeared stable without edema and only subtle pallor.

Fig. 2. Optic disk photographs and presentation showing severe, hemorrhagic bilateral optic disk edema OD and OS (a), and then resolution of disk edema 4 months after presentation (b).
Discussion

Fulminant IIH, which occurs in about 2% of IIH patients, can rapidly lead to irreversible vision loss and demands urgent ophthalmology and neurosurgical consultation [2]. Given the rarity of fulminant IIH, there have only been a handful of reports in the literature, with the largest study being by Thambisetty et al. [2] in 2007 with 16 patients. All patients in the Thambisetty et al. [2] study presented with a headache, and many also complained of nausea and vision changes. Additional symptoms that were reported included diplopia, vomiting, neck stiffness. Medical therapy with acetazolamide and methylprednisolone was initiated on every patient; however, no symptom improvement was seen, and they had all ultimately underwent surgical intervention. Except for the sex, the demographic and clinical presentation of our patient was in keeping with other documented presentations of fulminant IIH: female, elevated BMI, headache, and vision changes. Unlike our patient, all of these documented cases involved surgical interventions of optic nerve sheath fenestration, ventriculoperitoneal shunting, or lumboperitoneal shunting [2, 3]. There are cases in the literature of fulminant IIH treated without surgery. Espino Barros Palau et al. [4] reported a patient with fulminant IIH who had declined surgery and showed significant improvement with acetazolamide alone. However, the patient did undergo three serial lumbar punctures which may have contributed to the improvement. Nonetheless, the final visual fields were very good and comparable to our patient. Similarly, Shaia and Elzie [5], highlighted a case of fulminant IIH who was managed exclusively with acetazolamide for 1 year since she had experienced significant improvement within 5 days of treatment initiation. However, there was still a significant visual field defect at final follow-up with a mean deviation of −8.76 dB.

It remains unknown why some patients may improve significantly with only medical treatment. The diagnostic lumbar puncture may have been influential in the improvement in this case, and sufficient lowering of the CSF pressure may have reversed the distal transverse sinus stenosis and resulted in improvement in the CSF pressure. A repeat venogram was not available for us to determine if this was the case. A prolonged CSF leak may also occur in some patients. No significant weight loss occurred over the treatment course for this to be a possibility. If medical management is used as an initial treatment strategy as in this case, frequent and careful neuro-ophthalmology follow-up is required to ensure that there is no worsening of visual function. Surgical intervention may be required if this occurs. In this case, it remains unknown if his visual outcome would have been better if surgical intervention was undertaken. Detailed assessment of his visual outcome including contrast sensitivity and more detailed visual field testing was not performed as it is generally not part of routine care. It is possible that these parameters may have been better with surgical intervention. Nonetheless, this patient was very happy with his visual outcome and achieved normal visual acuity. Patients should be counselled on the role of surgery in the management of fulminant IIH and should be aware of the risks of deferring this for medical treatment as there are limited data in this patient population.

In conclusion, fulminant IIH is an uncommon form of the disease that results in rapid onset of symptoms. Most patient have significant visual deficits and require close follow-up. Although surgical management is often required, this case demonstrates that medical management may be sufficient for visual recovery and resolution of the disease.

Statement of Ethics

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. This study protocol was reviewed and approved by the University of Toronto Ethics Research Board. Case reports do not require approval number
by the Institutional Review Committee; therefore, this report granted an exemption from requiring an ethics approval number.

**Conflict of Interest Statement**

The authors have no conflicts of interest to declare.

**Funding Sources**

There were no funding sources for this study.

**Author Contributions**

Ojas Srivastava: design of the work, analysis, interpretation, and draft and revision for intellectual content. Jonathan A. Micieli: corresponding author, analysis, interpretation, revision for intellectual content, and final approval of the version to be published.

**Data Availability Statement**

Any data and additional information related to this study are available upon request through emailing the corresponding author. Data are available on request as it contains patient-specific confidential information.

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