

Grand Rounds

A 24-year-old woman with rapidly progressing vision loss

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History

A 24-year-old woman was referred to the Department of Ophthalmology, McMaster University, Hamilton, Ontario, for urgent neuro-ophthalmological evaluation following rapidly progressive vision loss and a 3-week history of constant, severe headache. The patient was otherwise healthy with no past medical or ophthalmic history. She had no previous history of such episodes, and she denied any history of recent trauma or infection. Her body mass index was 31, with a reported weight loss of 10 kg over the last 4 months. She was on oral contraceptives but denied taking any other medication.

Exam

On initial examination, the patient's best-corrected visual acuity was 20/100 in the right eye and 20/70 in the left eye. The pupils were symmetrically 3 mm in diameter and poorly responsive to light in both eyes, with no evidence of a relative afferent pupillary defect. Intraocular pressure was recorded as 16 mm Hg in each eye. Slit-lamp biomicroscopy was unremarkable. Dilated fundus examination revealed marked optic nerve head swelling suggestive of papilledema and tortuous and dilated retinal venules (Figure 1A).

Ancillary Testing

An initial head computed tomography (CT) scan was unremarkable. Lumbar puncture demonstrated an elevated opening pressure of 60 cm H₂O. The cerebrospinal fluid (CSF) composition was normal. Magnetic resonance imaging (MRI) of the head revealed increased perioptic space around both optic nerves with flattening of the posterior sclera suggestive of increased intracra-

nial pressure but was otherwise unremarkable. Magnetic resonance venography (MRV) was normal, with no evidence of dural sinus thrombosis. Visual field assessment (Humphrey 24-2) demonstrated severe and diffuse visual field loss bilaterally with preservation of central vision, with a mean deviation of -32.0 dB in the right eye and -28.6 dB in the left eye (Figure 2A; pattern deviation not shown due to severely depressed fields).

Treatment

At initial presentation to the emergency room, the patient's severity of vision loss prompted the patient's being admitted and placed on intravenous methylprednisolone 1 g daily for 3 days as well as oral acetazolamide 250 mg 4 times daily for 2 days. She was unable to tolerate a higher dose of acetazolamide. By the time of neuro-ophthalmological consultation, her visual symptoms had not improved, and she had new-onset abducens nerve palsy of the left eye with diplopia on left lateral gaze.

Given the rapidly progressive visual loss despite maximal tolerated medical treatment, a lumboperitoneal shunt surgery was urgently performed. Opening pressure at the time of the procedure was not recorded. All medications were subsequently discontinued.

One week after surgery, the patient's vision, diplopia, and headaches had improved. On examination, she had a best-corrected visual acuity of 20/25 in the right eye and 20/30 in the left eye. Pupils were equal and reactive. Ocular motility examination showed resolution of her abducens nerve palsy. Funduscopic examination showed improvement of the papilledema (Figure 1B), and visual field assessment exhibited improved visual fields bilat-

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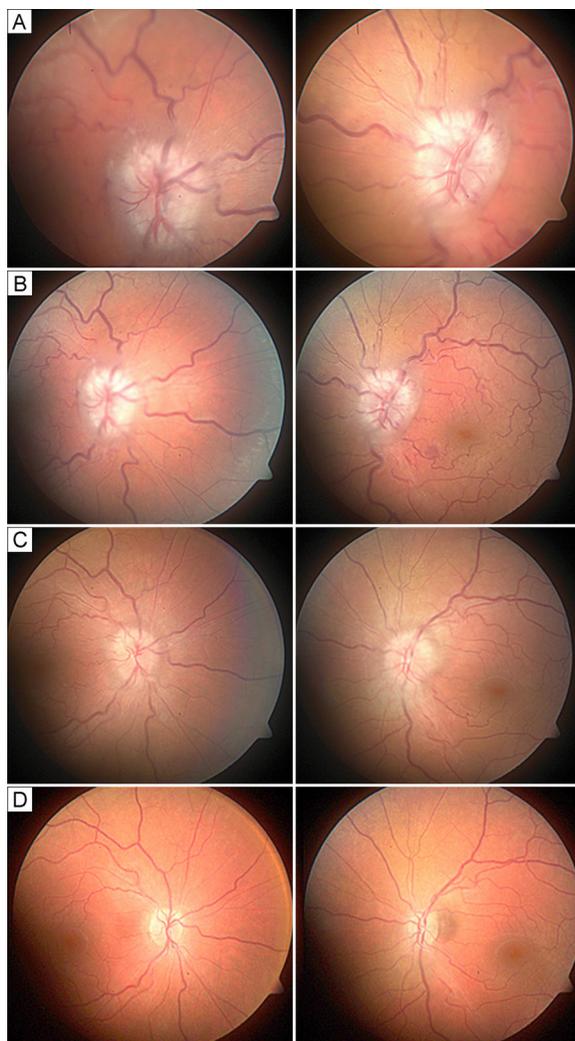


Figure 1. Fundus images of the right eye (left column) and left eye (right column) at acute presentation (A), and at 1 week (B), 6 weeks (C), and 18 months (D) after surgery.

erally, with a mean deviation of -24.4 dB in the right eye and -18.5 dB in the left eye. Postoperative follow-up at 1, 5, 9, 13, and 18 months demonstrated continued resolution of papilledema and retinal venular dilation and tortuosity (Figure 1C) with improvement of visual fields, with a mean deviation of -13.8 dB in the right eye and -8.4 dB in the left eye (Figure 2D). At final follow-up, 18 months postoperatively, visual acuity had stabilized at 20/25 in each eye. There was mild pallor of both optic nerves, suggesting permanent optic nerve damage. Visual field testing showed persistent inferonasal deficits bilaterally. On follow-up examination, there was mild peripapillary hyperpigmentation nasal to the left optic nerve but no evidence of choroidal neovascularization.

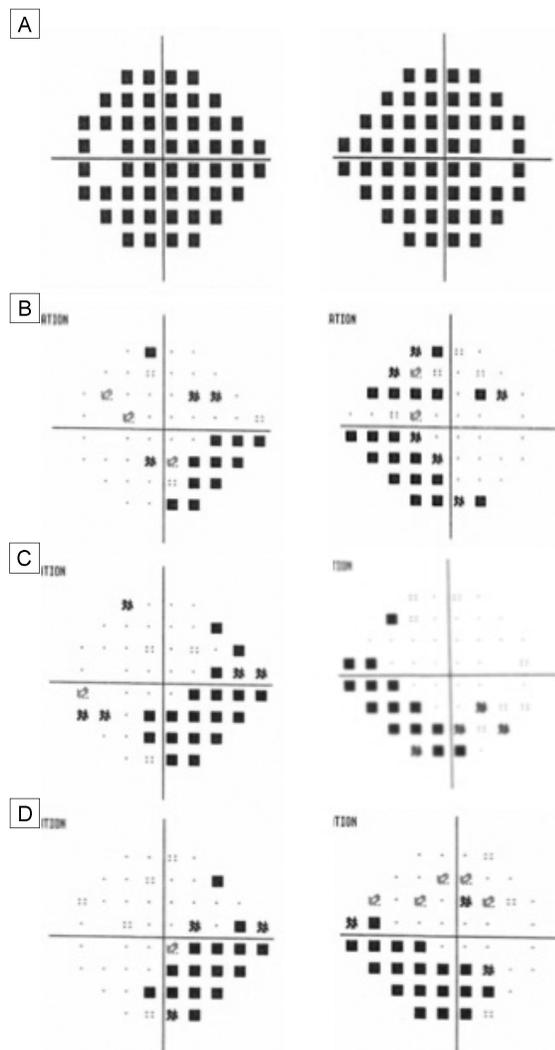


Figure 2. Humphrey's 24-2 visual field total and/or pattern deviation results of the right eye (right column) and left eye (left column). A, Total deviation at acute presentation (pattern deviations not shown due to severely depressed fields). B, Pattern deviation at 6 weeks after surgery. C, Pattern deviation at 4 months after surgery. D, Pattern deviation at 1 year after surgery.

Differential Diagnosis

Bilateral optic nerve head edema is presumed to be due to increased intracranial pressure (ICP) until proven otherwise. Other potential causes of bilateral optic nerve head swelling include malignant hypertension, intracranial tumor, hydrocephalus, and vascular abnormalities, such as dural sinus thrombosis. Pseudo-papilledema, including congenital drusen can also mimic the presentation of idiopathic intracranial hypertension (IIH). Other conditions to consider are bilateral anterior ischemic

optic neuropathy, optic nerve papillitis, or other infectious, inflammatory, or infiltrative etiologies affecting the optic nerves.

Diagnosis and Discussion

The elevated ICP without any clinical, laboratory, or radiographically identifiable etiology led to the diagnosis of IIH. The severity of visual loss and the rapidly progressive nature of this case reflected the fulminant form of IIH, prompting urgent diagnosis and management.

The overall incidence of IIH is 1–3 per 100,000, but it rises dramatically to 20 per 100,000 among female patients who are obese or have experienced recent weight change.^{1,2} Fulminant IIH is a particularly acute and rapidly progressive form of IIH, with an incidence of 2.2% to 2.9% of all new IIH cases.³

Normally, vision loss in IIH is secondary to chronic papilledema⁴; however, progressive vision loss is rare and may indicate secondary causes of increased ICP, such as a meningeal process or venous sinus thrombosis.^{5,6} Fulminant IIH is an acutely severe and rapidly progressive form of IIH, with resultant permanent visual sequelae.^{3,7–13} It is defined as acute onset of signs and symptoms of intracranial hypertension; (<4 weeks between onset of initial symptoms and severe visual loss), rapid worsening of vision loss over several days, and a normal MRI and MRV (or CT venogram).³ There is limited literature on fulminant IIH.^{8–12}

The pathophysiology of IIH remains unknown. Vision loss in papilledema is thought to be secondary to elevated CSF pressure transmitted to the anterior optic nerve sheath, resulting in axoplasmic flow stasis and subsequent intraneuronal ischemia.¹⁴ It is unclear why the condition manifests more rapidly and severely in some patients. The clinical characteristics of fulminant IIH are similar to those in the nonacute form.³

The treatment of fulminant IIH aims to alleviate symptoms and to preserve visual function. Fulminant IIH requires surgical intervention. Close observation and temporizing measures, such as repeat lumbar punctures, lumbar drain placement, acetazolamide and intravenous steroids are indicated prior to surgery.³ The patient in our case was treated with 1 g daily acetazolamide, because she was unable to tolerate a higher dose. Although patients can be treated with up to 4 g daily of acetazolamide, few patients can actually tolerate the side effect profile.

The use of corticosteroid therapy in the context of IIH has been suggested to decrease CSF formation at high ICP, to increase CSF absorption, and to mitigate papilledema independent of any change in ICP.¹⁵ Nevertheless, long-term use of steroids is not recommended due to well-established side effects.

Surgical management consists of CSF diversion procedures, namely, lumboperitoneal shunts and ventriculoperitoneal shunts, as well as optic nerve sheath fenestration (ONSF). In the United States, from 1988 to 2002, the frequency of CSF shunting procedures (lumboperitoneal and ventriculoperitoneal) for the treatment of IIH increased by 350%.¹⁶ Recently, venous sinus stenting has been performed on selected patients with IIH, although this remains an area of debate.¹⁷ To our knowledge, there have been no randomized, prospective trials undertaken to date that compare the efficacy and visual outcomes of these procedures in treating IIH.¹⁸ In addressing the underlying etiology of increased ICP, CSF diversion techniques improve papilledema and reduce further visual loss; however, they do not restore visual loss.

ONSF has also been shown to prevent visual deterioration and even improve visual function in some patients with IIH.¹⁹ However, up to one-third of patients with initially improved symptoms following ONSF experience worsening of visual acuity and visual fields over time. Thus, these patients require long-term follow-up, as deterioration of visual function may necessitate repeat procedures for ONSF failures. Overall, the decision to perform CSF diversion procedures or ONSF often depends on local availability and expertise as well as the predominance of the presenting symptoms. CSF shunting is often undertaken when headache is the primary presentation, whereas ONSF is carried out when vision loss is predominant.^{20,21}

Regardless of the choice of procedure, there is consensus for rapid and definitive treatment in rapidly progressive and refractory cases of IIH, as delay in surgical intervention can lead to worse visual outcomes.^{3,21} Thambisetty et al conducted the largest reported case series of fulminant IIH in 16 patients, who underwent surgical CSF shunting procedure or ONSF.³ Visual function improved in 14 of 16 patients postoperatively, although 8 patients (50%) remained legally blind at last follow-up; visual fields remained severely altered in all cases. Notably, the 8 patients who remained legally blind had a median delay in surgical intervention of 6.5 days from the time of diagnosis (range, 3–37 days), whereas those that regained significant visual function had a median delay of only 2 days (range, several hours

to 4 days), highlighting the importance of prompt and aggressive management.

Altogether, the acute progression and long-term visual sequelae of fulminant IHH in otherwise young healthy patients merits appropriate recognition, emergent neuro-ophthalmic evaluation, and possible surgical intervention. Surgical management is recommended in medically refractory cases with established progressive visual loss.

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