Fulminant idiopathic intracranial hypertension

Madhav Thambisetti, MD, PhD; Patrick J. Lavin, MD; Nancy J. Newman, MD; and Valérie Biousse, MD

Abstract—Objective: To describe the incidence and characteristics of acute and rapidly progressive visual loss in idiopathic intracranial hypertension (IIH). Methods: We reviewed the medical records of all patients with IIH seen at two institutions. “Fulminant IIH” was defined as the acute onset of symptoms and signs of intracranial hypertension (less than 4 weeks between onset of initial symptoms and severe visual loss), rapid worsening of visual loss over a few days, and normal brain MRI and MR venography (or CT venogram). Results: Sixteen cases with “fulminant IIH” were included (16 women, mean age 23.8 years [range 14 to 39 years]). All were obese. One patient had iron-deficiency anemia, four had systemic hypertension, and none had known sleep apnea syndrome. Acute or subacute headache, nausea and vomiting, and visual loss were present in all patients. The first lumbar puncture performed for the diagnosis showed a mean CSF opening pressure of 54.1 cm H2O (range 29 to 60 cm H2O). In addition to the initial lumbar puncture, medical treatment included acetazolamide (1 to 2 g/day) in all patients, and IV methylprednisolone in four patients. Repeat lumbar punctures were performed in 11 of the 16 patients. Surgical treatment (optic nerve sheath fenestration in five cases, lumboperitoneal CSF shunting procedure in nine cases, and ventriculoperitoneal shunting procedure in two cases) was performed because of ongoing visual loss in all cases. The median delay between evaluation in neuro-ophthalmology and surgery was 3 days (range a few hours to 37 days). All patients reported dramatic improvement of headaches and vomiting following surgery. Visual function improved in 14 cases, although 8 patients (50%) remained legally blind. Visual fields remained severely altered in all cases. Conclusion: Severe and rapidly progressive visual loss suggests “fulminant idiopathic intracranial hypertension” and should prompt aggressive management. Urgent surgery may be required in these patients, and temporizing measures such as repeat lumbar punctures, lumbar drainage, and IV steroids considered.

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Idiopathic intracranial hypertension (IIH) is increased intracranial pressure (ICP) with normal CSF contents, in the absence of an intracranial mass, hydrocephalus, or other identifiable cause.1,2 The major morbidity of IIH is progressive, insidious visual loss from chronic papilledema.1,3,9 Although progressive visual loss is common in poorly managed or noncompliant patients, acute presentation with rapidly progressive visual loss is rare and usually points to secondary causes of intracranial hypertension such as a meningeal process or venous sinus thrombosis.1,3,6,7 Rapid recognition of acute IIH, also described as “fulminant” or “malignant” IIH, is important, as it may prompt emergent surgical treatment.1,8,16 We present a series of 16 patients with “fulminant IIH” who developed early, severe, and rapidly worsening visual loss and evaluate the incidence of this disorder at two institutions.

Methods. The medical records of all cases with IIH seen in the Neuro-Ophthalmology Clinics at Emory University between 1996 and 2006 and Vanderbilt University between 2003 and 2006 were reviewed. Only cases with definite IIH according to the recently updated modified Dandy criteria2 were selected, including 1) symptoms and signs of generalized intracranial hypertension such as headache, papilledema, sixth nerve palsies; 2) documented elevated ICP; 3) normal CSF composition; 4) no evidence of hydrocephalus, mass, or structural or vascular lesion on brain MRI; specifically, no evidence of cerebral venous thrombosis. All IIH cases were reviewed in detail to identify patients with “fulminant IIH,” defined as follows: 1) acute onset of symptoms and signs of intracranial hypertension; 2) less than 4 weeks between onset of initial symptoms and severe visual loss; 3) rapid worsening of visual loss over a few days. Only patients who underwent brain MRI and MR venography (MRV) or CT venogram to rule out cerebral venous thrombosis were included. The study was approved by the Emory and Vanderbilt Institutional Review Boards. Patients’ characteristics were recorded, including age, gender, obesity (body mass index criteria), associated factors such as medications, systemic hypertension, anemia, and sleep apnea. Presenting features, visual acuity and visual fields, presence of transient visual obscurations, tinnitus and diplopia, time between the onset of symptoms and worst visual loss, time between the first lumbar puncture and surgery, and the median delay between evaluation in neuro-ophthalmology and surgery were recorded. Visual fields were assessed using standard automated perimetry. The study was approved by the institutional review boards of both institutions.

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From the Departments of Neurology (M.T., N.J.N., V.B.), Ophthalmology (N.J.N., V.B.), and Neurological Surgery (N.J.N.), Emory University, Atlanta, GA; Departments of Neurology (P.J.L.) and Ophthalmology (P.J.L.), Vanderbilt University, Nashville TN; and MRC Centre for Neurodegeneration Research (M.T.), Institute of Psychiatry, King’s College London, UK.

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Address correspondence and reprint requests to Dr. V. Biousse, Neuro-ophthalmology Unit, Emory Eye Center, 1365-B Clifton Rd. NE, Atlanta, GA 30322; e-mail: vbiousse@emory.edu

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onset of symptoms and surgical treatment, time between neuroophthalmic examination and surgical treatment, CSF opening pressure, treatment, and visual outcome were also recorded.

Results. A total of 483 cases with definite IIH were seen at Emory University between 1996 and 2006. Among these cases, 14 (2.9%) were diagnosed with “fulminant IIH.” Between 2003 and 2006, 89 cases with IIH were evaluated at Vanderbilt University, among which 2 (2.2%) were diagnosed with “fulminant IIH.” The clinical characteristics of these 16 consecutive patients seen in tertiary care centers with “fulminant IIH” are detailed in table E-1 on the Neurology Web site (www.neurology.org).

By definition, the 16 reported patients had normal brain MRI and MRV (or CT venogram) as well as normal CSF contents, thereby ruling out cerebral venous thrombosis or a meningeal process as the cause of their acute intracranial hypertension. None had empty sella. All cases were young women, either obese (14 cases) or overweight (2 cases). Mean age was 23.8 years (range 14 to 39 years). One patient had iron-deficiency anemia, and all other patients had a normal hematocrit. Systemic hypertension was present in four patients; none had accelerated or “malignant” systemic hypertension. None were known to have sleep apnea syndrome. Headache was a prominent presenting symptom in all patients and was accompanied in four patients by neck pain or stiffness or both and by nausea and recurrent vomiting in nine patients. Tinnitus was reported by nine patients, transient visual obscurations by eight patients, and diplopia by five patients. Decreased vision was a prominent complaint in all patients with central visual acuity impairment. Thirteen patients (81.2%) noted subacute visual loss at the same time they had headaches. Three patients had decreased vision a few days after the onset of severe headaches.

Severe loss of visual acuity and bilateral severe bilateral papilledema were noted at presentation in all cases (figure 1). Formal visual field testing, performed with Goldmann visual field testing or Humphrey visual field testing, showed severe bilateral constriction in all cases (figure 2). The first lumbar puncture performed for the diagnosis showed a mean CSF opening pressure of 54.1 cm H2O (range 29 to 70 cm H2O).

Visual loss was very rapidly progressive in all patients. The time between the patients’ first symptom and their worst visual loss ranged from 7 to 28 days (mean 16.1 days) (table E-1, column 2).

In addition to the diagnostic lumbar puncture, medical treatment included acetazolamide (1 to 2 g/day) in all patients and IV methylprednisolone in four patients. Although the initial lumbar puncture was followed by transient improvement in headaches, all patients experienced almost immediate recurrent headaches and persistent loss of vision. Hence, repeat lumbar punctures were performed in 11 of the 16 patients (6 cases had two lumbar punctures, 3 cases had three lumbar punctures, and 2 cases had four lumbar punctures). Repeat lumbar punctures resulted in only transient improvement of symptoms and signs, and surgical treatment (optic nerve sheath fenestration in five cases, lumboperitoneal CSF shunting procedure in nine patients, and ventriculoperitoneal shunting procedure in two patients) was performed because of ongoing visual loss. The median delay between evaluation in neuro-ophthalmology and surgery was 3 days (range a few hours to 37 days) (table E-1, column 7). All patients reported dramatic improvement of headaches and vomiting following surgery. Visual function improved in 14 cases, although 8 patients (50%) remained legally blind. Visual fields remained abnormal in all cases (table E-1). All 8 patients who were not legally blind at follow-up had undergone surgery within a few hours to 4 days (median 2 days) after being evaluated by us; in the 8 legally blind patients, surgery had been performed within 3 to 37 days (median 6.5 days) after neuro-ophthalmic evaluation.

Discussion. Although many reports emphasize the risk of visual loss in IIH, there are few prospective studies documenting the natural history of IIH. Typically, visual acuity is affected late in the course of IIH, and automated visual fields are required to detect insidious visual field constriction, which is

Figure 1. Initial optic nerve appearance of four patients with fulminant idiopathic intracranial hypertension (the right eye is on the left and the left eye is on the right). Note the severe bilateral disc edema with dilated veins, numerous exudates, and peripapillary hemorrhages.
present in a large majority of cases. Our series of 16 patients who presented with dramatic symptoms and signs of intracranial hypertension and developed rapid, devastating visual loss is obviously very different from the typical description of IIH. Because of this atypical presentation, each of these patients had a thorough workup ruling out a secondary cause of isolated raised ICP. Specifically, none of these patients had a meningeal process or cerebral venous thrombosis, which more commonly present with such dramatic symptoms and signs.

Except for the temporal profile and the severity of visual loss, the clinical characteristics of our patients did not differ from those with typical IIH. All our patients were young women, and all were obese or overweight. None had any acute illness or other factors that may have precipitated such explosive clinical symptoms and signs. One patient had long-standing iron-deficiency anemia, and four had relatively controlled systemic hypertension. Previous studies emphasized that poor prognostic factors for visual loss in IIH include long-standing swelling of the disk with atrophy, visual field or acuity loss at first examination, delay in treatment, systemic hypertension, age older than 40 years, male gender, and anemia. However, these criteria applied to patients with classic IIH who developed insidious, progressive visual loss. Although it is possible that our patients may have had long-standing papilledema without headaches prior to becoming aware of visual loss, the rapid onset of symptoms, the appearance of the disc edema, and the absence of empty sella on brain MRI suggested an acute process. The severe sudden and rapidly progressive visual loss is most likely related to axoplasmic stasis and optic nerve ischemia associated with a sudden rise in ICP. However, the pathophysiology of IIH remains unknown, and it is unclear why these patients had such an acute evolution of a characteristically subacute to chronic disorder.

Although several investigators have mentioned seeing patients with acute IIH, similar reports are exceedingly rare in the literature. In large series of patients, the prevalence of severe loss of visual acuity as a presenting feature of the illness is low. For example, in a series of 63 patients with IIH, only 5 cases had decreased visual acuity at the time of diagnosis. Of these, only one patient experienced an acute loss of central vision, which occurred after subtemporal surgical decompression. The remaining patients experienced deterioration of vision over a period of 2 to 4 months. Similarly, in a prospective study of 50 patients with newly diagnosed IIH, there was a low incidence of visual acuity loss as an early symptom of the condition. In a study of 57 patients with IIH, initial visual acuity loss was noted in only 9 patients, and most developed gradual worsening of vision in the months after the initial symptoms. Four well documented IIH cases presented with acute severe visual loss, which improved

Figure 2. Goldmann and Humphrey visual fields of the same four patients with fulminant idiopathic intracranial hypertension (the left eye is on the left and the right eye is on the right). The left column (A, C, E, G) shows the initial visual fields, and the right column (B, D, F, H) shows the final visual fields after treatment. Humphrey visual fields were performed on the 24-2 SITA Standard or Fast programs. The first two cases (A/B and C/D) are good examples of very limited improvement of visual function; these two patients are legally blind. The last two cases (E/F and G/H) are examples of improved visual function, but with constricted fields.
after urgent surgical treatment.\textsuperscript{10-12} Three other reports\textsuperscript{14,16} also described six IIH cases with subacute visual loss. Because both Emory and Vanderbilt are tertiary care centers with highly specialized neuro-ophtalmology services, we are more likely to evaluate IIH patients with unusual clinical presentations. Even in this setting, only 2.9\% of the Emory IIH patients and 2.2\% of the Vanderbilt IIH patients had fulminant IIH, emphasizing the rarity of this presentation. Nevertheless, although fulminant IIH is rare, it affects young, otherwise healthy women, who often become legally blind over the course of a few days. The need for emergent aggressive treatment of these patients is commonly overlooked by emergency room physicians and by neurologists, who are often the first to evaluate these patients when headaches and nausea are predominant and often more alarming than visual loss.\textsuperscript{9}

Although no controlled trial has evaluated surgical treatments in IIH, there is general agreement that surgery is indicated in cases of progressive visual loss.\textsuperscript{1,9} The type of procedure varies and may depend on local availability and expertise of neurosurgeons (who perform CSF shunting procedures) and ophthalmologists (who perform optic nerve sheath fenestrations). In institutions in which both procedures are readily available, a CSF shunting procedure is usually chosen in cases with severe headaches responding to lumbar punctures (sometimes after placement of a temporary lumbar drain); optic nerve sheath fenestrations are often preferred when severe visual loss associated with papilledema is present. The timing of surgery also varies widely, but in most cases, surgery is performed only when visual function deteriorates despite lumbar puncture and medical treatment, including diuretics and sometimes steroids.\textsuperscript{1,3,16} Our series of patients with fulminant IIH emphasizes the need for aggressive treatment in cases with such rapid visual loss; indeed, it has been emphasized that immediate surgery may result in partial improvement of visual function.\textsuperscript{10-12} Our study supports these findings. IV steroids may be helpful before surgery\textsuperscript{16} and were administered to four of our cases. Repeat lumbar punctures help decrease the ICP efficiently while waiting for surgery and were performed prior to referring the patients to our services in 11 cases. Placement of a lumbar drain may also be a temporizing measure while awaiting surgery. Our patients underwent surgery a median of 3 days after being evaluated by us. Only three patients, who were hesitant about undergoing surgery, delayed the procedure by 10, 12, and 37 days after their neuro-ophthalmic evaluation. These three patients remain legally blind. All the patients who had significant improvement of their visual function underwent surgery within a few hours to 4 days after neuro-ophthalmic evaluation.

It is important not to delay surgery in those rare patients with acute symptoms and signs of IIH and rapidly progressive visual loss. This decision is primarily based on the availability of surgeons able to perform a CSF shunting procedure or an optic nerve sheath fenestration within a few hours. Temporizing measures include repeat lumbar punctures, placement of a lumbar drain, and IV steroids, during which time patients are closely observed. Therefore, admission to the hospital is often preferred, as it accelerates the workup and appropriate treatment of these patients with fulminant IIH.

References

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