

Severe visual loss associated with idiopathic intracranial hypertension (IIH) in pregnancy

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Abstract: Idiopathic intracranial hypertension (IIH) in pregnancy is often responsive to conservative management and usually carries a favorable prognosis. Pregnant patients may be managed with dietary control, corticosteroid therapy, diuretics, and occasionally serial lumbar puncture. We report an unusual case of permanent visual loss due to IIH in pregnancy, requiring aggressive CSF diversion and bilateral optic nerve sheath fenestration after failure to improve with a brief course of medical management and a lumbar puncture. Despite resolution of the papilledema, the subject's acuity improved only to 20/40 in the right eye and remained no light perception in the left eye.

Keywords: idiopathic intracranial hypertension, pregnancy, visual loss

Introduction

Idiopathic intracranial hypertension (IIH) in pregnancy is often responsive to conservative management and usually carries a favorable prognosis. Pregnant patients may be managed with dietary control, corticosteroid therapy, diuretics, and occasionally serial lumbar puncture. The following case represents an unusual report of permanent visual loss due to IIH in pregnancy, which required aggressive cerebrospinal fluid diversion and bilateral optic nerve sheath fenestration.

Methods

Retrospective chart, radiographic, and photographic review.

Case report

A 22 year-old African American (G6P2, no spontaneous fetal loss) with a 13 week intrauterine pregnancy was transferred to our facility for further management of IIH. During her uncomplicated pregnancy, the patient gained approximately two pounds from a prepartum weight of 155 lbs. (prepartum body mass index = 28). The past medical history was otherwise unremarkable. A sister had systemic lupus erythematos. The patient took an oral contraceptive for six months prior to the pregnancy, and a daily prenatal vitamin during the pregnancy.

At gestational week 12, she presented to a local emergency department with complaints of headache and blurred vision for approximately one day. She was treated with acetaminophen/hydrocodone over the next six days. Despite treatment, the patient developed worsening headache and blurred vision, photophobia, dizziness, nausea, vomiting, and muscle stiffness and spasm in the neck and trapezius. She remained afebrile throughout her entire course. Her visual acuity at that time was reported as 20/100 in the right eye and light perception in the left eye. A computed tomographic (CT) scan without contrast and magnetic resonance venography (MRV), were both normal. She underwent a lumbar puncture in the lateral recumbent position with an opening pressure of 460 mmH₂O. Cerebrospinal fluid (CSF) studies, including cytology,

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Gram stain, culture, cryptococcus antigen, and stain for acid-fast bacilli, were normal (protein = 32; glucose = 69; leukocytes = 2; 92% lymphocytes; red blood cells [RBC] = 1). The patient was treated with 250 mg intravenous methylprednisolone succinate every six hours and ceftriaxone. Other hematological values at that time included hemoglobin of 12.6 gram/deciliter, hematocrit of 36.4%, leukocyte count of $10.4 \times 10^9/\text{liter}$ (L) and a platelet count of $274 \times 10^9/\text{L}$. A serum rapid plasma reagin was nonreactive.

The patient was transferred to our facility for further management. Best corrected visual acuities were 20/100 in the right eye and no light perception (NLP) in the left eye. The patient correctly identified 10/10 Ishihara color plates with the right eye. The left pupil failed to react to light, although the efferent response was intact. Humphrey 24-2 fastpac visual field testing of the right eye revealed a mean deviation of -16.1 . There was an enlarged blind spot, dense nasal depression, and peripheral constriction (Figure 1). Extraocular movements were full, and the patient was orthophoric by Hirschberg testing. Slit lamp examination was noncontributory. Dilated funduscopic examination revealed moderately severe, bilateral papilledema without evidence of vitritis or other retinal disturbances (Figure 2).

The patient was admitted to the high-risk obstetrics service with the diagnosis of IIH. Review of previous imaging studies (CT, MRV), and a subsequent high quality magnetic resonance image of the brain and orbits with

fat suppression and contrast at our facility, revealed no abnormalities. Specifically, there was no hydrocephalus, mass lesion, meningeal or optic nerve enhancement. Her systolic blood pressure ranged from 104 to 137 mm of mercury (mmHg) and the diastolic from 55 to 60 mmHg.

Additional studies included: Westergren erythrocyte sedimentation rate (ESR) = 62 mm/hour (hr); normal thyroid function panel; protein C = 175 (normal range: 60–140); activated partial thromboplastin time = 23.7 seconds, and an equivocal Lyme screen. Relevant coagulation measures, including homocysteine level, protein S activity, Dilute Russell viper venom, and anticardiolipin IgG and IgM, were all normal. C-reactive protein and antinuclear antibody were normal. In addition, the patient had negative serologies for human immunodeficiency virus and hepatitis C. A tuberculosis skin test was negative. A hemoglobin electrophoresis was normal.

The patient received a gram of methylprednisolone succinate per day in divided doses. One day after admission, she underwent a bilateral ONSF and concurrent placement of a lumbar drain (5 ml/hr). Microscopic evaluation of specimens of the right and left optic nerve sheaths was normal. Gram stain and culture of the CSF bag specimen obtained during drainage were unremarkable, and repeated analysis revealed only rare lymphocytes, 31 RBC, and normal protein and glucose levels. The CSF VDRL was negative. The lumbar drain was removed on post-operative day three and the patient was discharged home on post-operative day four on 60 mg of prednisone.

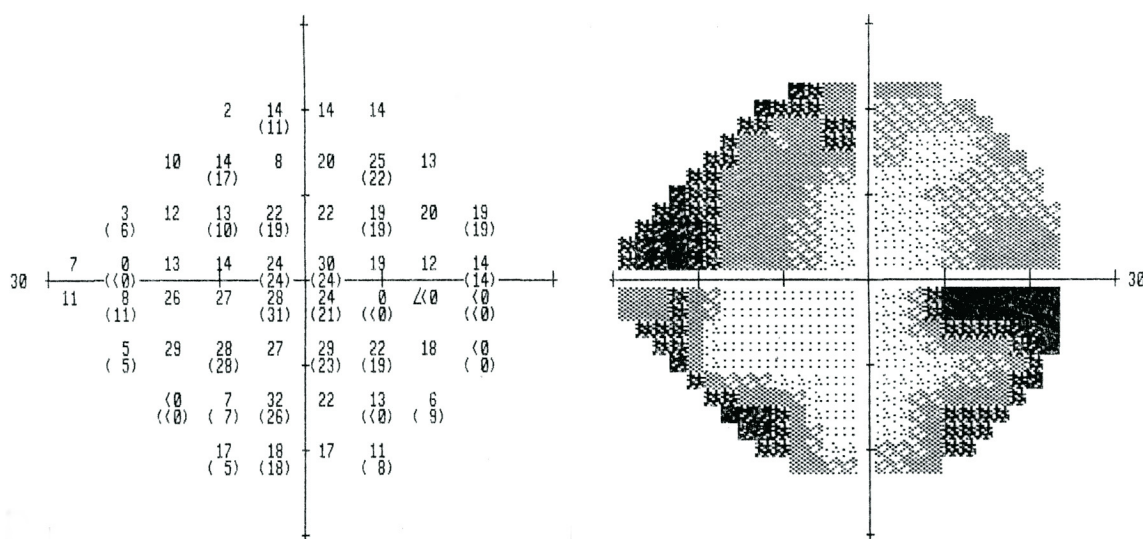


Figure 1 Humphrey 24-2 visual field testing with mean deviation of -16.1 ; right eye.



Figure 2 Severe disc edema, splinter hemorrhage, venous tortuosity, nerve fiber layer opacification and cotton-wool spots; **A**, right eye; **B**, left eye.

In follow-up, the visual acuity in the right eye improved to 20/40 and remained NLP in the left eye. The patient's funduscopic examination showed resolution of disc edema but pallor developed bilaterally (Figure 3).

Discussion

The patient's course and tempo of visual loss is unusual for IIH. Extensive hematological, serologic, and structural evaluation for other causes of elevated intracranial

pressure was unrevealing. Specifically, there was no evidence of meningitis, inflammatory optic neuropathy nor venous sinus thrombosis. Three laboratory abnormalities deserve further discussion. An elevated ESR was felt to be secondary to pregnancy rather than due to a primary inflammatory or viral process. A recent study by van den Broe and colleagues (2001) concluded that ESR ranges for pregnant patients vary with gestational age and hemoglobin level. For anemic patients in the first half of

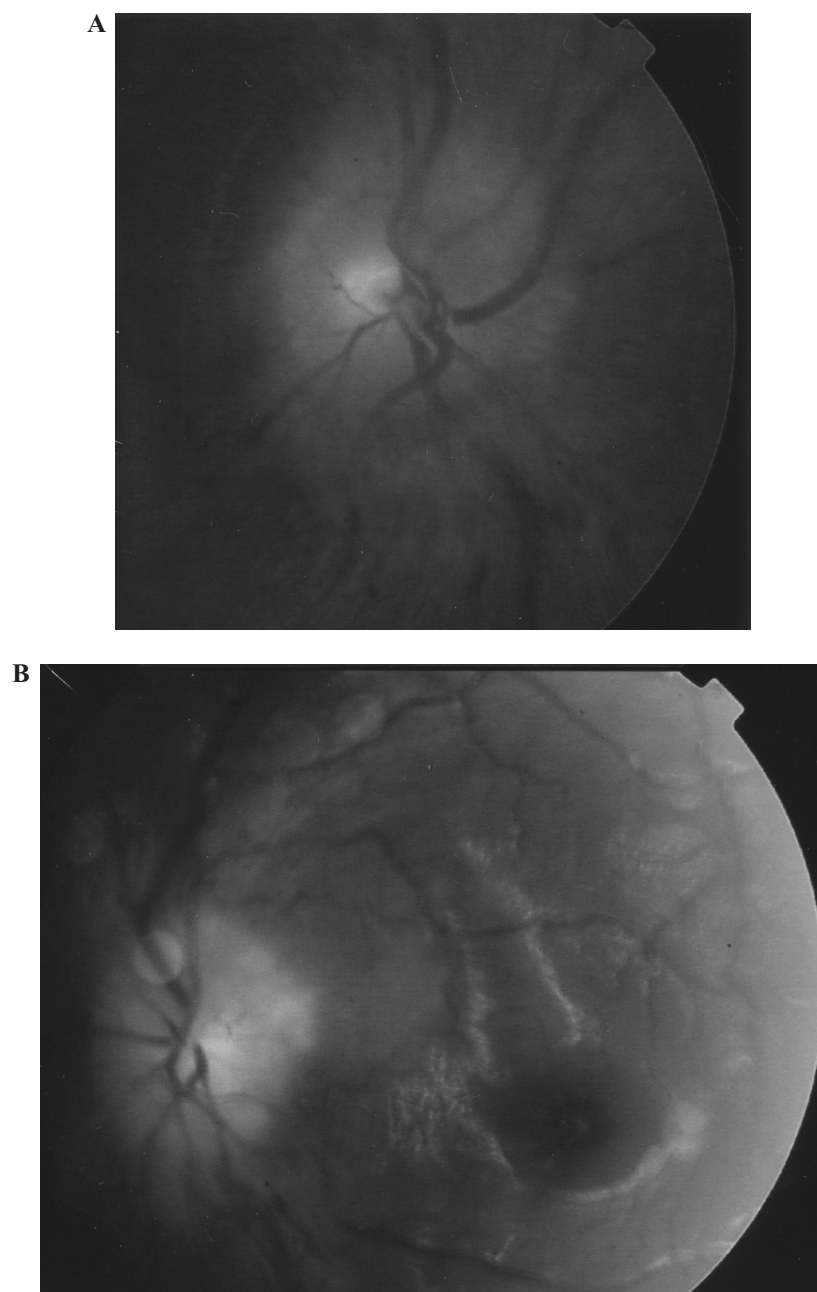


Figure 3 Significant decrease of papilledema three days following ONSF and placement of lumbar drain; **A**, right eye; **B**, left eye.

pregnancy, ESR is expected to be in the range of 40 to 95 mm per hour. Thus, the ESR was within normal limits for the patient's stage of pregnancy. Secondly, according to a prospective study by Oruc and colleagues (2000), Protein C and Protein S anti-coagulant proteins typically decrease during a normal pregnancy on a dilutional basis. It is possible that the modest elevation in protein C in this case represents an acute phase response. Finally, the equivocal Lyme testing did not meet the Center for

Disease Control and Prevention criteria for the diagnosis of Lyme disease.

More than 70 cases (Nickerson and Kirk 1965; Elia et al 1968; Powell 1972; Caroscio and Pellmar 1978; Shekleton et al 1980; Anderson 1983; Kassam et al 1983; Koontz et al 1983; Digre et al 1984; Peterson and Kelly 1985; Katz et al 1989; Shapiro et al 1995; Evans and Friedman 2000; Huna-Baron and Kupersmith 2001) of IIH during pregnancy are reported in the English language since 1960. A review of

these reports revealed no case of blindness and only a handful of reported cases of permanent visual field loss (Nickerson and Kirk 1965; Powell 1972; Shekleton et al 1980; Shapiro et al 1995). Our patient was unique in that she not only experienced permanent loss of visual acuity and visual field, but the onset and magnitude of loss was both rapid and extreme. This type of clinical progression of IIH in pregnancy is not typically reported. Furthermore, based on the cases presented in the literature, IIH in pregnancy is often responsive to conservative management with diet, bed rest, steroids, diuretics or lumbar puncture(s) (Nickerson and Kirk 1965; Powell 1972; Caroscio and Pellmar 1978; Alderson 1983; Kassam et al 1983; Koontz et al 1983; Digre et al 1984; Peterson and Kelly 1985; Katz et al 1989; Huna-Baron and Kupersmith 2001). Only a minority of cases requires aggressive therapy involving surgical intervention. In our case, conservative medical management failed and carbonic anhydrase inhibitors were not used because of their relative contraindication in pregnancy (category C: animal studies show adverse fetal effects[s]; no controlled human studies). Thus, rapid surgical intervention was needed to halt the progressive visual loss and probably contributed to ultimate improvement in visual acuity and field in the right eye.

Disclosure

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References

- Alderson TL. 1983. Nonrecurrent pseudotumor cerebri in pregnancy: report of case. *J Am Osteopath Assoc*, 82:728–31.
- Caroscio JT, Pellmar M. 1978. Pseudotumor cerebri: occurrence during the third trimester of pregnancy. *Mt Sinai J Med*, 45:539–41.
- Digre KB, Varner MW, Corbett JJ. 1984. Pseudotumor cerebri and pregnancy. *Neurology* 34:721–9.
- Elia M, Ben-Tovim N, Bechar M, et al. 1968. Recurrent benign intracranial hypertension (pseudotumor cerebri) during pregnancy. *Obstet Gynecol*, 31:685–8.
- Evans RW, Friedman DI. 2000. The management of pseudotumor cerebri during pregnancy. *Headache*, 40:495–7.
- Huna-Baron R, Kupersmith MJ. 2001. Idiopathic intracranial hypertension in pregnancy. *J Neurol*, 249:1078–81.
- Kassam SH, Hadi HA, Fadel HE, et al. 1983. Benign intracranial hypertension in pregnancy: current diagnostic and therapeutic approach. *Obstet Gynecol Surv*, 38:314–21.
- Katz VL, Peterson R, Cefalo RC. 1989. Pseudotumor cerebri and pregnancy. *Am J Perinatol*, 6:442–5.
- Koontz WL, Herbert WNP, Cefalo RC. 1983. Pseudotumor cerebri in pregnancy. *Obstet Gynecol*, 62:324–7.
- Nickerson CW, Kirk RF. 1965. Recurrent pseudotumor cerebri in pregnancy. Report of 2 cases. *Obstet Gynecol*, 26:811–13.
- Oruc S, Saruc M, Koyuncu FM, et al. 2000. Changes in the plasma activities of protein C and protein S during pregnancy. *Aust N Z J Obstet Gynaecol*, 40:448–50.
- Peterson CM, Kelly JV. 1985. Pseudotumor cerebri in pregnancy. Case reports and review of literature. *Obstet Gynecol Surv*, 40:323–9.
- Powell JL. 1972. Pseudotumor cerebri and pregnancy. *Obstet Gynecol*, 40:713–18.
- Shapiro S, Yee R, Brown H. 1995. Surgical management of pseudotumor cerebri in pregnancy: case report. *Neurosurgery*, 37:829–31.
- Shekleton P, Fidler J, Grimwade J. 1980. A case of benign intracranial hypertension in pregnancy. *BJOG*, 87:345–7.
- van den Broe NR, Letsky EA. 2001. Pregnancy and the erythrocyte sedimentation rate. *BJOG*, 108:1164–7.

