Fulminant Idiopathic Intracranial Hypertension in Pregnancy

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Abstract
Fulminant IIH in pregnancy requires multidisciplinary collaboration and immediate CSF diversion.

Introduction
Fulminant idiopathic intracranial hypertension (IIH) is a rare, acute, rapidly progressive form of idiopathic IIH. Its unique occurrence during pregnancy posed challenges beyond simply treating the visual loss.

Case Report
We report a patient with fulminant idiopathic IIH during pregnancy and describe the issues we faced. In just under 4 weeks, this 28-year-old pregnant woman developed holoccephalic pulsing headache, photophobia, vomiting, pulse-synchronous tinnitus, blurred vision in the left eye, and binocular oblique diplopia.

Examination showed weight of 100 kg; height of 1.55 m; body mass index of 42 kg/m\textsuperscript{2}; blood pressure of 130/75; visual acuities of 20/40 on the right and 20/200 on the left; color
vision of 6/17 Ishihara plates on the right and 1/17 on the left; a left relative afferent pupillary defect; bilateral grade 3–4 Frisén optic disc edema; left peripapillary hemorrhages; left greater than right eye abduction deficits; and otherwise normal neurological examination. She was immediately admitted to hospital for investigation and management.

Optical coherence tomography showed average retinal nerve fiber layer thicknesses of 211 μm on the right and 180 μm on the left. Automated 24-2 perimetry showed mean deviation of −26.01 dB and concentric constriction in the right eye and mean deviation of −36.67 dB and generalized depression, denser nasally, in the left eye (Fig. 1).

Abdominal ultrasound showed a 13-week pregnancy. Magnetic resonance imaging (MRI) showed findings of IIH. Time-of-flight MRI venography showed stenoses of the superior sagittal and bilateral transverse and sigmoid sinuses and the right jugular vein.

Cerebrospinal fluid (CSF) opening pressure was 43 cm H₂O, and its contents were normal. A temporary lumbar drain was inserted, acetazolamide 3,000 mg daily begun, and a right occipital ventriculoperitoneal (VP) shunt inserted, after which the acetazolamide was tapered and discontinued.

Six months after the VP shunt operation, by mutual agreement of the high-risk obstetrician and the neurosurgeon, the patient delivered a healthy child by cesarean section. At 8 months, the examination showed visual acuities of 20/20-2 on the right and 20/70 on the left; color vision of 13/17 Ishihara plates on the right and 6/17 on the left; a left relative afferent pupillary defect; mild left optic disc pallor; and normal ocular ductions.

Visual fields showed mean deviation of −8.47 dB in the right eye and mean deviation of −17.23 dB in the left eye. There were left greater than right eye inferior nasal steps, inferior arcuate nerve fiber layer defect in the right eye, and inferior and superior arcuate nerve fiber layer defects in the left eye (Fig. 2). There have since been weight reduction to 85 kg and no shunt failure or relapse of the IIH.

**Discussion/Conclusion**

This patient fulfilled the criteria for fulminant IIH: acute onset of symptoms and signs of IIH; less than 4 weeks between onset of initial symptoms and severe visual loss; rapid worsening of visual loss over a few days [1, 2]. Fulminant IIH is rare to begin with [2], but the situation of fulminant IIH during pregnancy was unique and prompted multidisciplinary discussions and decisions about management.
Our overall approach to this challenge was (a) to adhere to principles of IIH management as set out in guidelines [3, 4] and of fulminant IIH management as far as they could be determined given its rarity [2] and (b) to adapt measures to her pregnant state. Weight reduction is the cornerstone of IIH treatment but could not be immediately implemented due to the pregnancy and because it would take too long to work given the urgency of the visual loss.

We were mindful of using acetazolamide in pregnancy; however, considering the risks and benefits, we noted that “there is no convincing evidence for an adverse effect for acetazolamide use in human pregnancy…” [5]. Acetazolamide alone, without CSF diversion [6], has reportedly been successful in fulminant IIH, but given the degree of visual dysfunction and the urgency we faced, we did not risk this treatment [2].

In general, the need for rapid effect in fulminant IIH renders the use of drugs alone impracticable. Topiramate should not be used in pregnancy because of its teratogenic risk [3]. Due to the lack of high class evidence, corticosteroids are not recommended for fulminant IIH [3] except perhaps, as alluded to below, as a temporizing measure pending definitive treatment. Adding furosemide to the acetazolamide was precluded by limited data.

The somatostatin analogue octreotide and 11b-hydroxysteroid dehydrogenase type 1 blocking agents are not yet being used in IIH. Consensus guidelines do not support the use of serial lumbar punctures [7].

Despite reports of success of venous sinus stenting in fulminant IIH [8], we avoided it because during our patient’s pregnancy there would be need for intraprocedural anticoagulation and for dual antiplatelet agents. The choices among surgical interventions in IIH remain controversial as they depend on local resources and practices and still lack evidence [9]. The Surgical Idiopathic Intracranial Hypertension Treatment trial was terminated due to low enrolment [10]. Surgical options for treating fulminant IIH include CSF diversion procedures (ventriculoperitoneal (VP) shunting or lumbarperitoneal shunting) and optic nerve sheath fenestration (ONSF). There are no prospective studies directly comparing the efficacy and complications of these interventions in patients with fulminant IIH [2]. The choices involve institutional familiarity with and availability of these procedures.

With respect to ONSF, local experience was relatively limited, it was unavailable in our tertiary hospital, and it could not be arranged in a timely fashion. The literature on ONSF is observational, mainly case series based, and largely retrospective. Risks include persistently raised intracranial pressure, vision loss, and diplopia. Worsening vision after a period of stabilization occurs in 34% of patients at 1 year and 45% at 3 years [3].
Various CSF diversion procedures were considered. Notwithstanding injunctions to avoid shunting in pregnancy [11], a VP shunt was felt to be the most straightforward procedure, with a lower revision rate than lumboperitoneal shunting [12]. In addition, unlike the seldom used lumboperitoneal, ventriculopleural, ventriculoatrial, lumbopleural, or lumboatrial shunts, VP shunts do not require X-rays for placement. A concern about VP shunting was the possibility that increased abdominal pressure during pregnancy could impair drainage of CSF. In practice, neither the consultant neurosurgeon nor the high-risk obstetrician had encountered that complication nor had they encountered peritonitis, another potential risk.

In the pregnant patient with IIH in general and in the pregnant patient with fulminant IIH in particular, multidisciplinary communication among relevant experienced clinicians should occur throughout pregnancy, peridelivery, and in the postpartum period [3]. Also, no specific mode of delivery should be chosen on the basis of the diagnosis of IIH alone [3]. The principles of managing fulminant IIH in pregnancy consist of the following steps in collaboration with obstetrics: (1) identify fulminant IIH based on the diagnostic criteria above; (2) admit the patient to hospital to confirm the diagnosis; consult neurology, ophthalmology, neuro-ophthalmology (if available), neurosurgery, and obstetrics; order MRI brain and orbits and MR venogram but without contrast because of the pregnancy; perform lumbar puncture; begin acetazolamide 2–4 g per day; consider intravenous methylprednisolone as a limited temporizing measure only with obstetrical approval but prioritize insertion of a temporizing lumbar drain; (3) within 4 days of diagnosis, undertake intervention in the form of VP shunt or ONSF but avoid venous sinus stenting because it would require X-rays and dual antiplatelet agents during pregnancy [2].

Statement of Ethics

Ethical approval was not required for this study in accordance with local guidelines. Our report does not contain any identifying information about the subject patient. Written informed consent was obtained from the patient for publication of the details of their medical case and the accompanying figures.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Conception and design (Felix Tyndel and Richard Wennberg); data collection (Felix Tyndel; Claude Steriade; and Antonio Gallo); manuscript preparation (Felix Tyndel and Richard Wennberg); critical appraisal (Felix Tyndel; Richard Wennberg; and Antonio Gallo); review of the manuscript (Felix Tyndel; Richard Wennberg; Claude Steriade; Antonio Gallo; and Ivan Radovanovic); final approval and agreement to be accountable for all aspects (Felix Tyndel; Richard Wennberg; Claude Steriade; Antonio Gallo; and Ivan Radovanovic).
Data Availability Statement

All the data can be found in the case report itself. Further inquiries can be directed to the corresponding author.

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