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A delicate balance: Managing intracranial hypertension in pregnancy

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Abstract

Idiopathic intracranial hypertension (IIH) is a condition marked by increased intracranial pressure with no identifiable cause, most commonly affecting young, obese women. We present the case of a 24-year-old primigravida with a body mass index of 37, ventricular septal defect repair, and type II diabetes mellitus, who presented at 15 weeks of gestation with blurred vision and reduced visual fields in her right eye. On examination, a grade 2 relative afferent pupillary defect was noted in the right eye, alongside bilateral optic disc swelling. Despite normal neuroimaging and cerebrospinal fluid analysis, a high opening pressure of 33.5 cmH_2O was documented during lumbar puncture. A multidisciplinary team from neurology, ophthalmology, and obstetrics, initiated treatment with acetazolamide and topiramate, along with a lumbar drain and serial lumbar punctures. The patient successfully delivered a healthy baby girl via emergency caesarean section at 37 weeks of gestation. After delivery, her optic nerve functions remained stable, although some chronic damage was evident. This case underscores the importance of multidisciplinary collaboration and careful treatment planning for IIH during pregnancy to safeguard both maternal and foetal health.

Keywords: Acetazolamide, cerebrospinal fluid diversion, idiopathic intracranial hypertension, pregnancy

INTRODUCTION

Idiopathic intracranial hypertension (IIH), also known as pseudotumour cerebri, is characterised by elevated intracranial pressure (ICP) without discernible aetiology. It predominantly affects young, obese women. Although relatively uncommon, it exhibits a prevalence of approximately 5% during pregnancy, often demanding comprehensive management due to the intricate interplay between maternal well-being and foetal health.[1] We present a case of IIH in pregnancy, highlighting the intricate management challenges posed by this unique clinical scenario.

CASE REPORT

A 34-year-old primigravida with type 2 diabetes mellitus and obesity (body mass index: 37kg/m^2), presented at 15 weeks of gestation with painless progressive blurred vision in her right eye, with reduced peripheral visual field. There were no double vision, headache, nausea, vomiting, and other neurological symptoms.

On examination, the visual acuity (VA) of the right eye was 4/60 and the left was 6/12. She also had grade 2 relative afferent pupillary defect (RAPD), reduced light brightness and red saturation in the right eye. Fundus examination revealed bilateral optic disc swelling. (Figure 1) Humphrey's visual field (HVF) assessment showed a constricted right visual field. Magnetic resonance imaging (MRI) of the arterial and venous system of the brain was normal. A lumbar puncture (LP) was performed and confirmed an elevated opening pressure of 33.5 cmH_2O, with no leucocytes, protein of 0.71g/L, and glucose level of 5.3 mmol/L. Oligoclonal band was not detected in the cerebrospinal fluid (CSF). Aquaporin-4 receptor antibody and myelin oligodendrocyte glycoprotein (MOG) antibody were negative. Connective tissue

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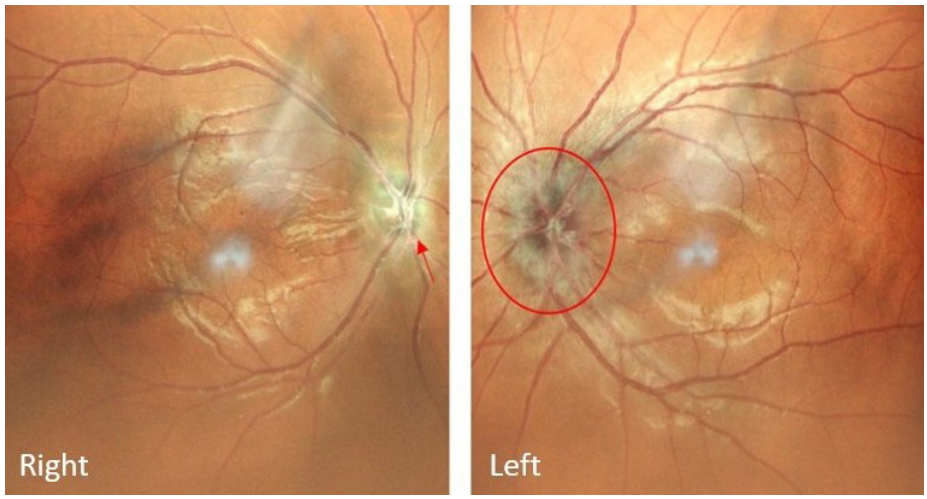

Figure 1. Right eye blurred disc margin seen (as in arrow) and left eye generalised optic disc swelling (as in circle).

disorder screen was unremarkable. A diagnosis of IIH was made based on her risk factor profile (obesity, pregnancy), raised intracranial pressure with normal neuroimaging and CSF analysis.

She booked for antenatal care at 9 weeks of gestation and was scheduled for follow-up in the high-risk clinic due to her history of ventricular septal defect repair and underlying type 2 diabetes mellitus. At the time of booking, she did not report any eye symptom. Her pregnancy was managed under the maternal-foetal medicine unit in collaboration with the cardiologist, neurologist, and endocrinologist. The foetus was slightly larger than expected for gestational age. Given her multiple comorbidities, she was planned for an elective caesarean section.

Given the sight-threatening effect of IIH, a lumbar drain was inserted and was followed by an improvement of her visual acuity where her best corrected VA was 6/18 on the right eye and 6/9 on the left eye. She was discharged after 14 days of hospitalisation and managed with a combination of weekly LP at daycare and oral topiramate 50mg twice daily. Subsequent eye follow-up at 18 weeks of gestation showed the VA of the right eye improved to 6/12 and the left eye to 6/6, despite persistent optic nerve dysfunction. Although the optic disc swelling eventually resolved, the right eye showed pallor of the optic disc indicating sign of chronic damage. Serial two-weekly optical coherence tomography (OCT RNFL) showed thinning of the right eye's retinal nerve fibre layer over time. (Figure 2)

Despite periodic LP with fluctuating opening pressures, ranging from 21-29 cmH_2O and topiramate, the ICP increased to 39 cmH_2O at 24 weeks of gestation. The treatment strategy was revised after a multidisciplinary team discussion to add acetazolamide at 250mg twice daily to her management plan of periodic LP and topiramate.

Her pregnancy progressed well; however, at 37 weeks, she underwent an emergency caesarean section due to reduced foetal movements.

She returned to the eye clinic 10 months post-delivery with a right eye VA of 6/12 with improvement in light brightness, red saturation and visual field test and left eye maintained good vision of 6/6. OCT RNFL showed persistent thinning of the nerve fibre layers of the right eye and resolution of optic disc swelling over the left eye.

DISCUSSION

Symptomatically, IIH often manifests with elevated ICP symptoms, with headache being the commonest symptom.[1,2] Some patients may solely present with ocular symptoms such as visual obscuration, double vision and loss of vision in severe cases.[2] The presenting symptoms and visual outcome were indifferent between pregnant and non-pregnant women.[1] A detailed eye examination is essential for signs of intracranial hypertension such as RAPD, binocular diplopia and extraocular movement impairment secondary to sixth or third cranial nerve palsies.[2] Papilloedema, which is the hallmark of intracranial hypertension, is almost always present in the early phase although there were also cases of IIH without papilledema (IIHWOP) reported. Although the exact cause of IIHWOP was unknown, there was an association

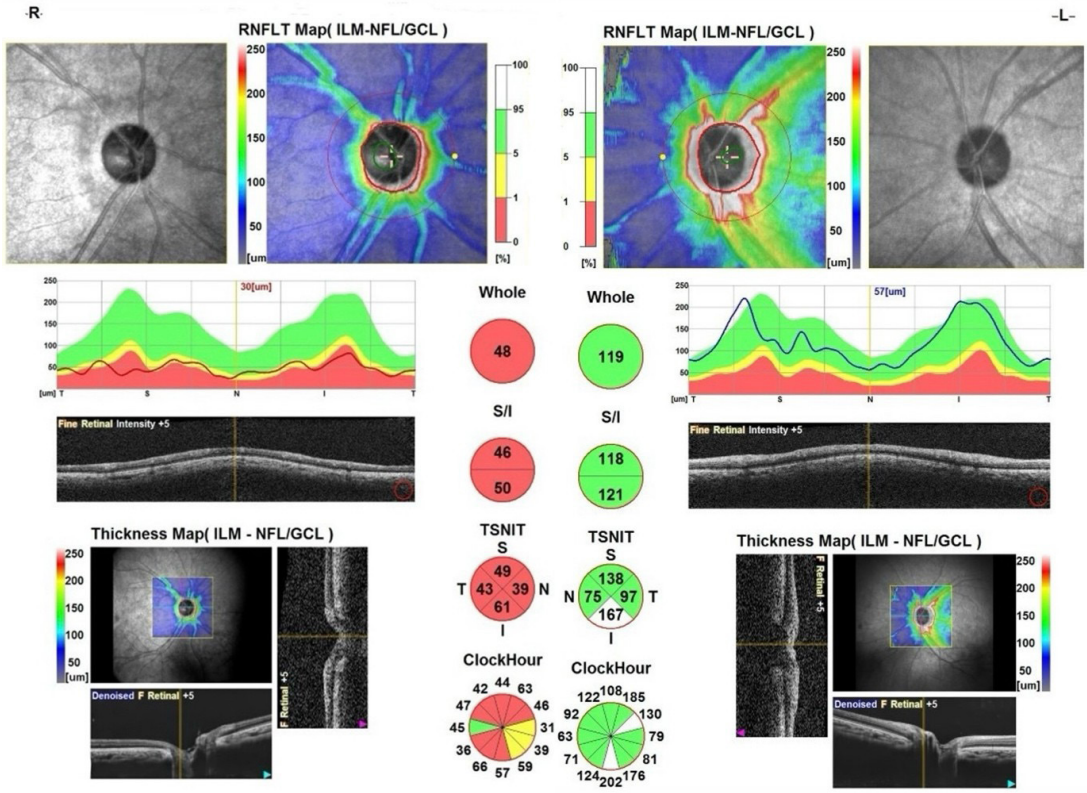

Figure 2. OCT RNFL shows RNFL thinning on the right eye and thickening on the left eye.

between bilateral transverse sinus stenosis with patients who present with chronic tension-type headache (CTTH) in IIHWOP.[3]

Treatment aims to preserve vision by reducing ICP. Weight loss, acetazolamide, and topiramate are the first-line treatments. However, according to Malaysian National Pharmaceutical Regulatory Agency (NPRA), acetazolamide is contraindicated in the first trimester. If to be started during the second and third trimester, potential benefits must justify the potential risk and careful evaluation of the foetus is needed before the initiation of acetazolamide as acetazolamide can exert teratogenic effects by inducing an acidotic embryonic environment.[4,5] However, no adequate studies have been done on humans. In a study conducted by Lee *et al.*, none of the patients had an adverse pregnancy outcome with no major or minor congenital malformations.[5] Topiramate use in pregnancy carries risks of congenital malformations, as well as preterm labour and premature delivery. The most frequent adverse events of topiramate that were reported to Malaysian NPRA were weight loss which could help in the management of IIH, drug inefficiency, dizziness, and stutter.[6]

In patients with refractory IIH with signs of deterioration of vision, surgical treatments can be considered which include optic nerve sheath fenestration (ONSF), lumbar drain, and CSF diversion such as lumboperitoneal shunt or VP shunt, and venous sinus stenting. Both VP shunt and LP shunt attempt to shunt fluid from the CSF to the peritoneal cavity to relieve elevated ICP; however, LP shunt has higher revision rates and risk of shunt obstruction as the pregnancy progresses and the uterus increases in size.[7] In our case, the patient showed clinical improvement following a lumbar drain insertion. Based on this response, and after a thorough discussion, she has opted for regular lumbar punctures instead of undergoing a shunt at this time.

CSF diversion can result in 66.8% improvement of both visual fields and visual acuity and resolution of papilledema was noted in 78.9% of patients who underwent CSF diversion surgery.[5] However, it is associated with a wide range of complications. Around 9.8% of these patients had serious complications, with shunt infection being the most common.[8]

The discordant improvement between visual acuity and opening pressure from LP resulted

in challenging management. Acetazolamide was started after considering the benefits that outweighed the risks. The role of VP shunt was discussed; however, given the risks of infection and the position of the shunt in the peritoneum that may be difficult in a pregnant woman, it was not deemed a practical approach.

A multidisciplinary approach involving obstetricians, neuro-ophthalmologists, neurologists, and neurosurgeons is important in managing IIH in pregnancy as it remains a challenge to balance the benefit of treatment to the patient and the effect on the foetus. For pregnant women with IIH, medical approaches such as weight management should be emphasized. If the condition threatens vision and causes significant headaches, medical options such as topiramate and acetazolamide can be offered to reduce CSF pressure. Surgical options can be considered once all other options are exhausted. While medical and surgical options exist, individualized care remains crucial to optimise outcomes.

DISCLOSURE

Ethics: Written informed consent was obtained from the patient for publication of the details of her medical case and any accompanying images.

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Conflict of interest: None

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