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Treatment of Asymptomatic Idiopathic Intracranial Hypertension

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Abstract:

Introduction: Idiopathic intracranial hypertension is a neuro-metabolic disorder comprising elevated cerebrospinal fluid pressure of unknown cause, which occurs commonly in women with obesity. Idiopathic intracranial hypertension is frequently diagnosed due to increased awareness and knowledge of magnetic resonance imaging findings. The first approach in such a situation is to rule out pseudo papilledema, to confirm the findings on magnetic resonance imaging, and to further confirm with lumbar puncture. Limited literature is available regarding the presentation and prognosis of asymptomatic idiopathic intracranial hypertension. Papilloedema may be incidentally observed during routine fundus examination in patients with symptoms.

Case report: This case report describes the clinical presentation, diagnostic evaluation, and management of a 21-year-old female with asymptomatic idiopathic intracranial hypertension. The patient was found incidentally with bilateral papilledema during a regular health check-up, prompting a thorough investigation to rule out secondary causes of elevated intracranial pressure. The patient was initially managed with topiramate for 6 months. A resolving trend of papilledema was seen during the follow-up period of 3 months.

Conclusions: The report highlights the challenges in managing asymptomatic idiopathic intracranial hypertension, emphasising the importance of a multidisciplinary approach for optimal patient care.

Key words: haemorrhage, topiramate, idiopathic intracranial hypertension (IIH), optical coherence tomography (OCT), papilloedema.

Idiopathic intracranial hypertension (IIH) is a neuro-metabolic disorder characterized by an unexplained rise in cerebrospinal fluid pressure, often seen in obese women. This neurological condition is identified by elevated intracranial pressure without a clear underlying cause. Idiopathic intracranial hypertension is frequently encountered nowadays, thanks to increased awareness and knowledge of magnetic resonance imaging (MRI) findings. The first approach in such a situation is to rule out pseudo-papilledema to confirm the findings on MRI and to further confirm it with lumbar puncture. There is a lack of literature regarding the clinical features and prognosis of IIH. Papilledema may be found incidentally during routine fundoscopic examinations in patients who show symptoms. This case report describes the clinical presentation, diagnostic evaluation, and management of a 21-year-old female with asymptomatic IIH. The patient was incidentally found to have bilateral papilledema during a regular health check-up, despite a lack of symptoms. This prompted a thorough investigation to rule out secondary causes of elevated intracranial pressure. The patient was managed initially with topiramate for 6 months. A resolving trend of papilledema was observed during the follow-up period of 3 months. The report highlights the challenges in managing asymptomatic IIH, emphasizing the importance of a multidisciplinary approach for optimal patient care.

Introduction

Idiopathic intracranial hypertension is characterized by increased intracranial pressure (ICP), resulting in chronic headaches, vision problems, and cognitive impairments. The occurrence of optic disc swelling, either directly or indirectly, plays a role in the visual decline linked to IIH. Studies show that the severity of papilledema is associated with the degree of visual loss [1]. This condition predominantly affects women of reproductive age who are also obese, highlighting the importance of regular screening for papilledema in this population [2, 3].

Case report

Patients who present with headaches and visual disturbances are frequently referred to ophthalmology and neurology clinics after incidental findings of papilledema are discovered during routine neurological evaluations. Upon further questioning, many of these patients report experiencing symptoms. Neuroimaging signs of increased intracranial pressure include conditions such as empty sella syndrome, lateral sinus collapse marked by smooth-walled venous stenoses, flattened globes, and fully unfolded optic nerve sheaths.

A 21-year-old woman was found to have incidental papilledema on an annual check-up. She had experienced no symptoms whatsoever involving the central nervous system or the eye for the preceding year and more. She was seen again by us in the department of ophthalmology and referred to neurology on confirmation of ophthalmic findings of papilledema. Her fundus picture showed almost bilateral symmetrical disc edema grade 2 (as per the modified Frisén scale) with nasal border elevation, with circumferential halo, without any major vessel obscuration, without any disc hemorrhage, and without any lumpy appearance of disc drusen (to rule out pseudo-papilledema). Her optical coherence tomography (OCT) showed increased average retinal nerve fiber layer thickness in both eyes (117 μm right eye, 120 μm left eye). She also underwent an automated visual field test, which did not reveal any blind spot enlargement and showed only a few nonspecific changes. On inquiry again, she did not reveal any headaches, giddiness, tinnitus, or visual disturbances. There was no history of intake of hormonal pills.

On examination, her blood pressure was 126/72. Her height was 157 cm, weight 79 kg, and body mass index (BMI) 32.05 kg/m^2.

She was investigated with a plain and contrast MRI of the brain, which revealed mild tortuosity of bilateral optic nerves with prominent perioptic spaces and hypoplastic left transverse and sigmoid sinuses (Fig. 1).

At presentation, fundus showed disc edema with fuzzy margins in both eyes, and this disc edema was resolved in subsequent follow-up (Fig. 2).

At case presentation, OCT showed increased average retinal nerve fiber layer in both eyes. It started to resolve during the follow-up period of 3 months (Fig. 3).

She underwent lumbar puncture in the lateral position, which revealed an opening pressure of 360 mm of the cerebrospinal fluid (CSF). Her CSF analysis was normal: CSF protein 17.4 mg/dL, cell count 3/μL, CSF sugar 61 mg/mL.

Her blood investigation revealed low hemoglobin at 10.5 g/dL, with mean corpuscular volume of 64.3 fL. Ferritin and iron were low at 2.9 ng/mL (10-291 ng/mL) and 1.1 μmol/L (9.0 to 30.4 μmol/L), respectively.

Since she had unequivocal evidence of papilledema and raised intracranial hypertension, she was started on treatment after

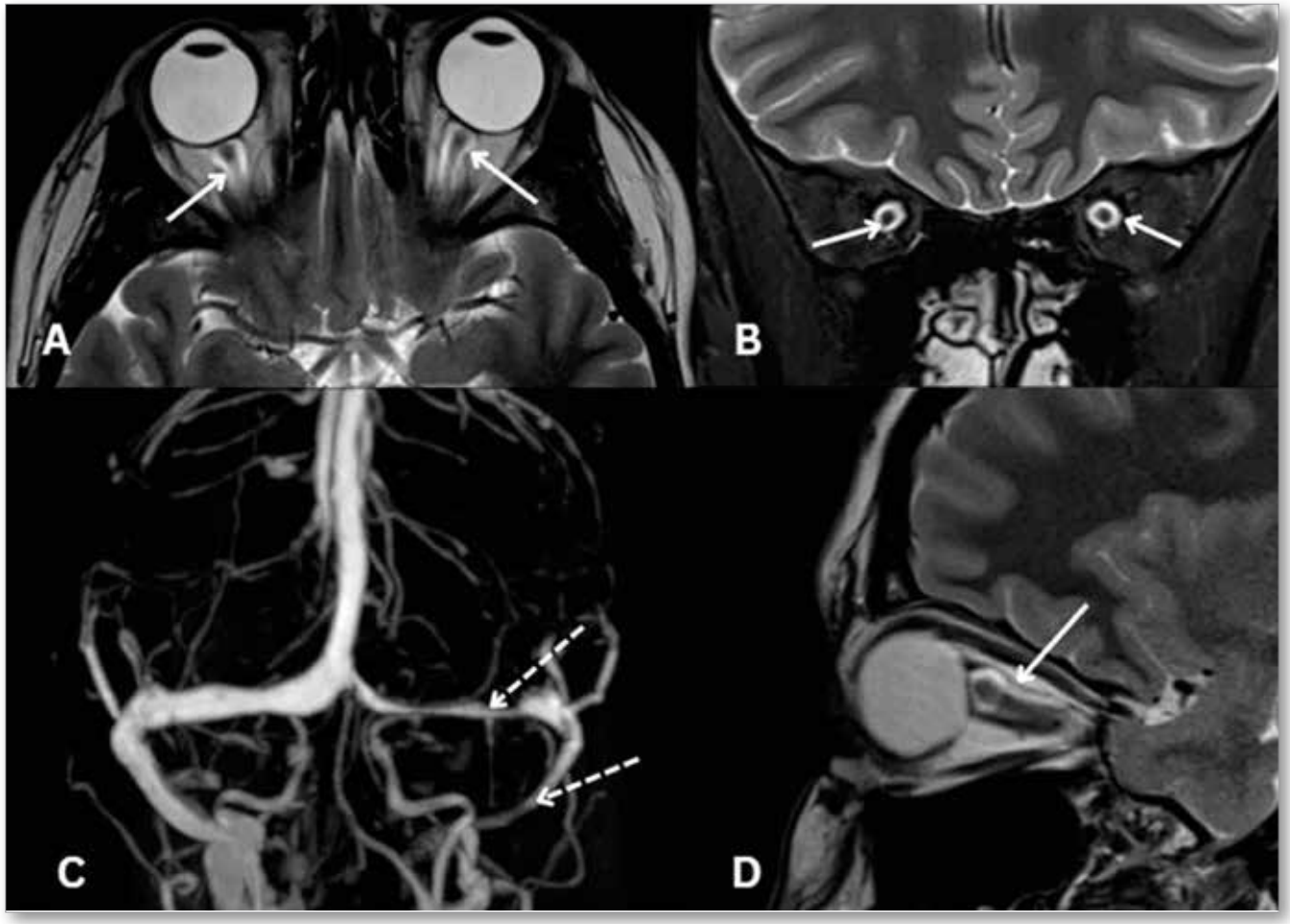

Fig. 1. Axial (A), sagittal (D) T2 and coronal T2FS (B) showing dilated bilateral perioptic spaces (solid arrows). MR venogram (C) shows stenosis of left transverse and sigmoid sinuses (dotted arrow).

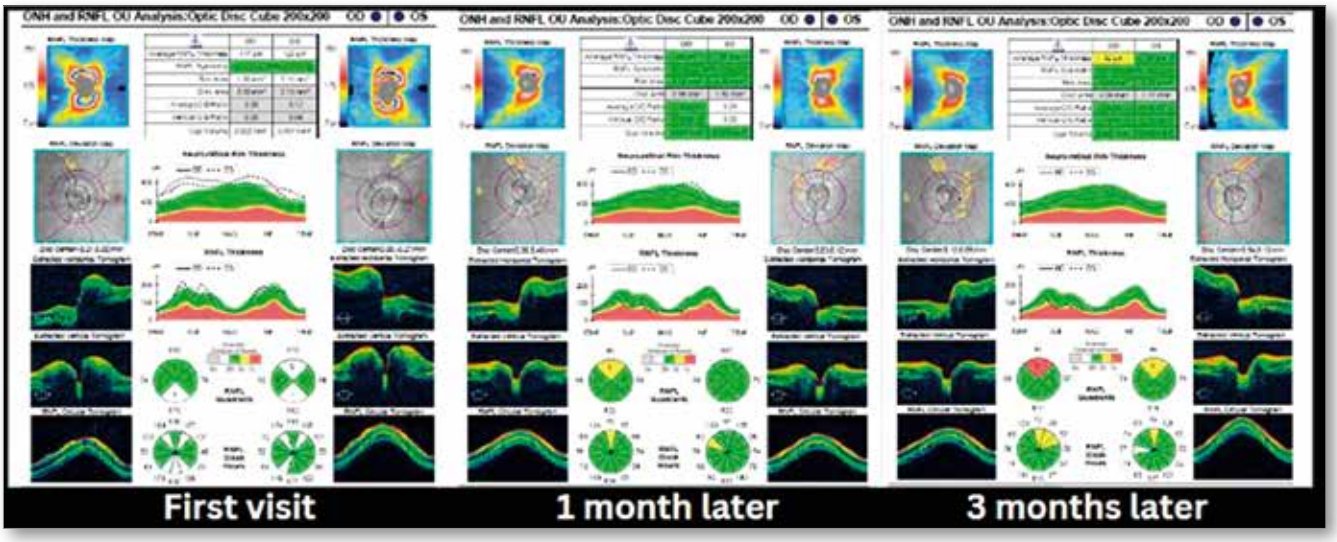

Fig. 2. Optical coherence tomography. Increased average retinal nerve fiber layer of both eyes at first visit, which showed a resolving trend over the follow-up period of 3 months.

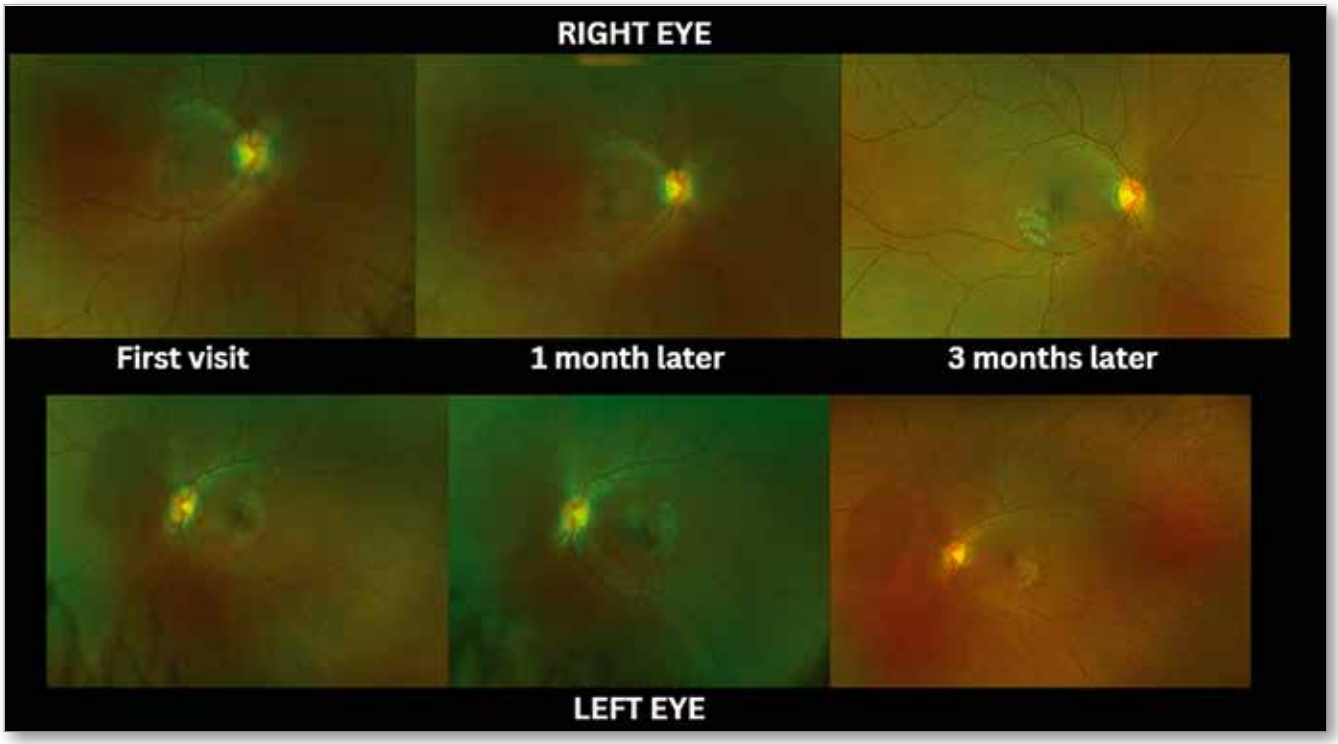

Fig. 3. Wide field fundus photos showing disc edema with fuzzy margins at first visit in both eyes. Disc edema was observed to be resolving in subsequent follow-up.

a detailed discussion and explanation to the patient. Since her BMI was high, it was decided to start her on topiramate 75 mg (Topamax, Janssen-Cilag GmbH, Johnson & Johnson, Platz 1, 41470 Neuss, Germany) per day (increased gradually) after explaining the possible side effects. She was started on topiramate 75 mg per day. One month after starting the medication, she complained of occasional headaches.

Three months after starting medication, she had lost 7 kg, and optical coherence tomography showed a resolving trend. Over the follow-up period of 3 months, her papilledema resolved. Her topiramate dosage was tapered to 50 mg per day, with the advice to maintain the same dose for the next three months. It is planned to stop topiramate 6 months after initiation and to keep her under regular follow-up.

Discussion

Thaller et al. in a longitudinal study of asymptomatic patients with IIH observed that all the patients would become symptomatic later [4].

With this presumption, the patient was started on therapy. Since her BMI was high, she was started on topiramate and improved significantly.

Many patients in the study by Vosoughi et al. [5] were detected incidentally, which again highlights the need for increased awareness of this condition. The clinical presentation could be variable and could change during the disease [6].

Though headache is the commonest presentation in IIH, tinnitus can be the only presentation [7].

The majority of children present with headaches, and IIH is an important condition in this population to be ruled out based on the correlation of history and MRI findings. Asymptomatic IIH can be a challenge in both adults and children when it is incidentally detected [7].

Treatment options include weight reduction, acetazolamide, topiramate, and surgical interventions (venous stent placement, thecoperitoneal shunt, and optic nerve fenestration) [8]. There is an urgent need for prospective cohort studies to explore the clinical progression of IIH. While papilledema is often discovered incidentally, true asymptomatic IIH is relatively uncommon when specifically investigated. The existence of asymptomatic disease does not seem to influence visual outcomes – either positively or negatively – compared to symptomatic cases. Diagnosing the worsening of papilledema in asymptomatic patients presents considerable challenges, indicating that this group may need ongoing ophthalmic assessments throughout their lives [9].

Obesity is consistently associated with IIH, likely reflecting an underlying metabolic disorder. Although achieving weight loss is notably difficult, it remains the only recognized intervention that can change the disease course in IIH. This case study revealed that variations in BMI were the most significant factor impacting visual outcomes. An increase in BMI during follow-up was linked to a decrease in OCT retinal nerve fiber layer (RNFL) and perimetry mean deviation (PMD), highlighting the importance of weight management for patients with IIH.

Headaches are a common symptom in IIH, with a gradual reduction in headache frequency noted over time. Two main factors contribute to the higher frequency of headaches and poorer prognosis at baseline. Future studies on IIH-related headaches must consider a history of migraines and daily headaches, as these elements can significantly affect clinical trial outcomes. Additionally, the introduction of calcitonin gene-related peptide (CGRP) therapy may influence headache outcomes in IIH in the future [10–12].

Medications used to lower ICP include acetazolamide, topiramate, and various diuretics. Analgesics are commonly used by some patients, especially those who may be taking multiple types of these medications. It is important to note that patients suffering from headaches on more than 15 days each month often show higher usage of analgesics. Future studies should include a control group to better understand the long-term effects related to IIH.

Incidental observation of papilledema was made during the check-up of a patient without any symptoms. Fundoscopy showed bilateral symmetrical disc edema grade 2 with nasal border elevation, with circumferential halo, without any major vessel

obscuration, without any disc hemorrhage, and without any lumpy appearance of disc drusen. Brain MRI revealed mild tortuosity of bilateral optic nerves with prominent perioptic spaces and hypoplastic left transverse and sigmoid sinuses. The patient was managed initially with topiramate 75 mg (Topamax, Janssen-Cilag GmbH, Johnson & Johnson, Platz 1, 41470 Neuss, Germany) per day for 3 months. Topiramate dosage was tapered to 50 mg per day, with the advice to maintain the same dose for the next 3 months, and it was planned to stop topiramate after 6 months. Optical coherence tomography showed a resolving trend of papilledema during the follow-up period of 3 months. Treatment of asymptomatic IIH may prevent permanent visual impairment. The majority of asymptomatic patients would eventually progress to symptomatic IIH.

Disclosure
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