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Bilateral Papilledema in Idiopathic Intracranial Hypertension: A Case Report



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ABSTRACT

Introduction: Idiopathic intracranial hypertension (IIH) or known as pseudo tumor cerebri is a challenging condition with raised intracranial pressure (ICP) in the absence of an identifiable cause. IIH occurs predominantly in women, and it has a striking association with obesity. The majority of patients presenting with IIH have symptoms that include a headache and visual disturbances. IIH presents challenges in management. This case report aims to discuss the clinical signs and management of idiopathic intracranial hypertension.

Case Report: A female 21 years old presented with visual field disturbances complaints since one week before admitted to the Hospital. Her symptom progressively worsened over time with headaches. Blood pressure was 150/80 mmHg, body weight was

110 kg, height was 167 cm, and body mass index was 40 kg/m². Visual acuity in both eyes was 6/6. The intraocular pressure was 17 mmHg on the right eye and 18 mmHg on the left eye. There was a constriction of vision's peripheral field and enlarged blind spot with Humphrey 24-2 visual field test. Fundus examination with optical coherence tomography of retinal nerve fiber layer revealed bilateral papilledema. The patient was diagnosed with papilledema et causa Idiopathic Intracranial Hypertension in both eyes. The patient was treated with acetazolamide and combination therapy with the neurology department.

Conclusion: IIH is a challenging disease with a collaborative department. Early and aggressive management is required to prevent irreversible visual impairment.

Keywords: Idiopathic Intracranial Hypertension, Obesity, Headaches, Acetazolamide

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INTRODUCTION

Idiopathic intracranial hypertension (IIH), previously known as pseudotumor cerebri, is a disorder characterized by elevated intracranial pressure (ICP) and remains a diagnosis of exclusion.¹ IIH occurs predominantly in women, is associated with obesity.² There are no underlying etiology combination, are accepted criteria for the diagnosis of IIH.³

IIH has a general population incidence of 0.5-2/100,000 and a higher incidence among women of childbearing age (12-20 per 100,000), usually between the ages of 25 and 36.⁴ IIH is ten times more prevalent in women than in men, and obesity increases the risk of developing IIH nearly 20-fold.⁴ In the last decade (2002-2014), the average age-adjusted and gender-adjusted annual incidence increased and was reported to be 2.4 per 100,000.⁵

In the absence of any intracranial findings, the clinical diagnosis of IIH is based on a series of classic signs and symptoms; however, the etiology of IIH remains elusive and mostly theoretical. Most patients with IIH have multiple symptoms

(headaches, transient visual obscurations, pulsatile tinnitus, back pain, dizziness, neck pain, visual blurring, cognitive disturbances, radicular pain, and typically horizontal diplopia). Investigation and management relies on symptoms and signs and needs an interdisciplinary team approach.^{2,6,7}

Thus, many treatment options or recommendations are driven by these theories, including those related to abnormal CSF physiology, such as increased production and/or decreased absorption, and pressure differentials within the venous sinus system.^{8,9} This report will discuss the clinical signs and management of idiopathic intracranial hypertension.

CASE ILLUSTRATION

This was a single case study and has approved by Ramata Eye Hospital Denpasar. Patient was followed up from June 2020 until September 2020.

A female 21 years old presented with visual field disturbances complaints on both eyes one week ago before being admitted to the Ramata Eye Hospital on June 24, 2020. Her symptom progressively

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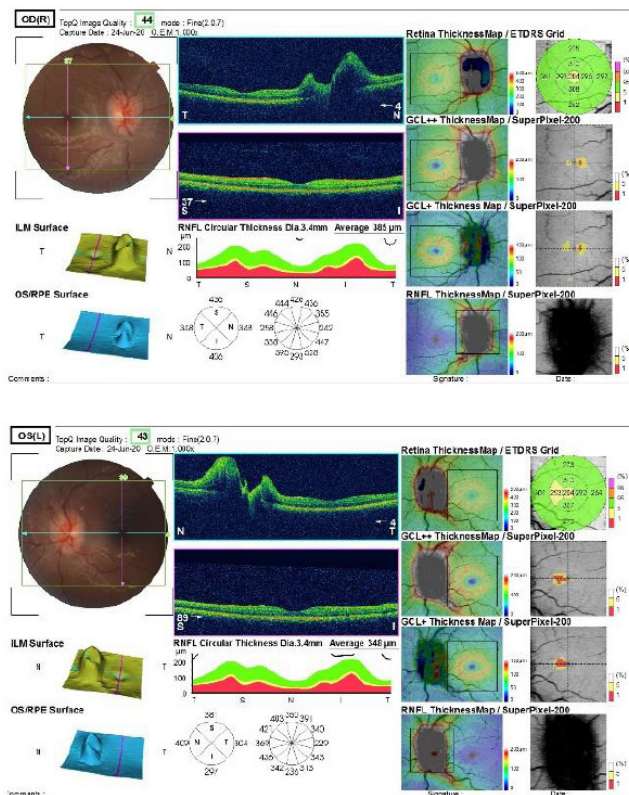


Figure 1. Bilateral papilledema was seen with OCT

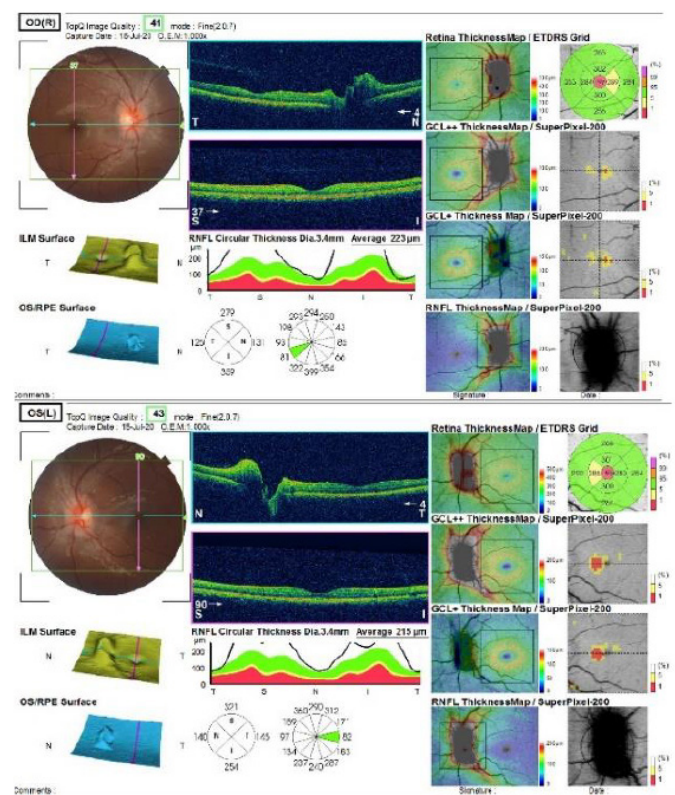


Figure 2. Bilateral papilledema was reduced with OCT

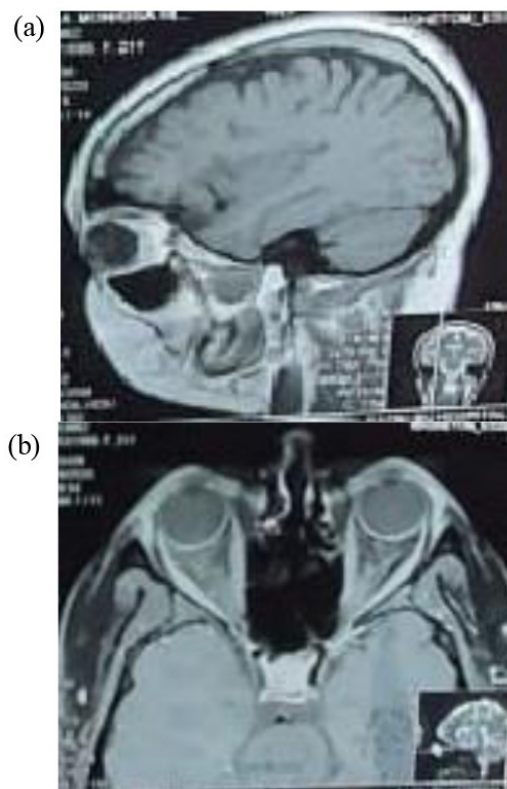


Figure 3. Neuroimaging inpatient patient with IIH. (a) Sagittal brain MRI with contrast show no abnormal lesions in the supra sella area. (b) Axial orbital MRI with contrast show no abnormalities

worsened over time with headaches. There was no associated pain, watering, or redness of the eye. There was no history of tinnitus, nausea or vomiting, dysphagia, fever, steroids, and accidents.

On examination found that the blood pressure (BP) 150/80 mmHg, weight 110 kg, height 167 cm, and body mass index (BMI) 40 kg/m². Visual acuity (VA) on both eyes was 6/6. Intraocular pressure (IOP) were 17 mmHg on the right eye (RE) and 18 mmHg on the left eye (LE). Anterior segment examination and extraocular movements were regular. There was a constriction of vision's peripheral field and enlarged blind spot with Humphrey 24-2 visual field test. Fundus examination with optical coherence tomography (OCT) revealed bilateral papilledema (**Figure 1**). The patient has been diagnosed with papilledema et causa Idiopathic Intracranial Hypertension (IIH) in both eyes. The patient was planned for brain and orbital magnetic resonance imaging (MRI) and consul to the neurology department. Patient treated with acetazolamide, neurotropic and neuroprotective agents. Patients are advised to lose weight with regular exercise and a balanced diet to achieve a normal BMI.

Patient-controlled on July 2020, the VA was 6/6 RLE and IOP 19 mmHg RE and 15 mmHg LE. The

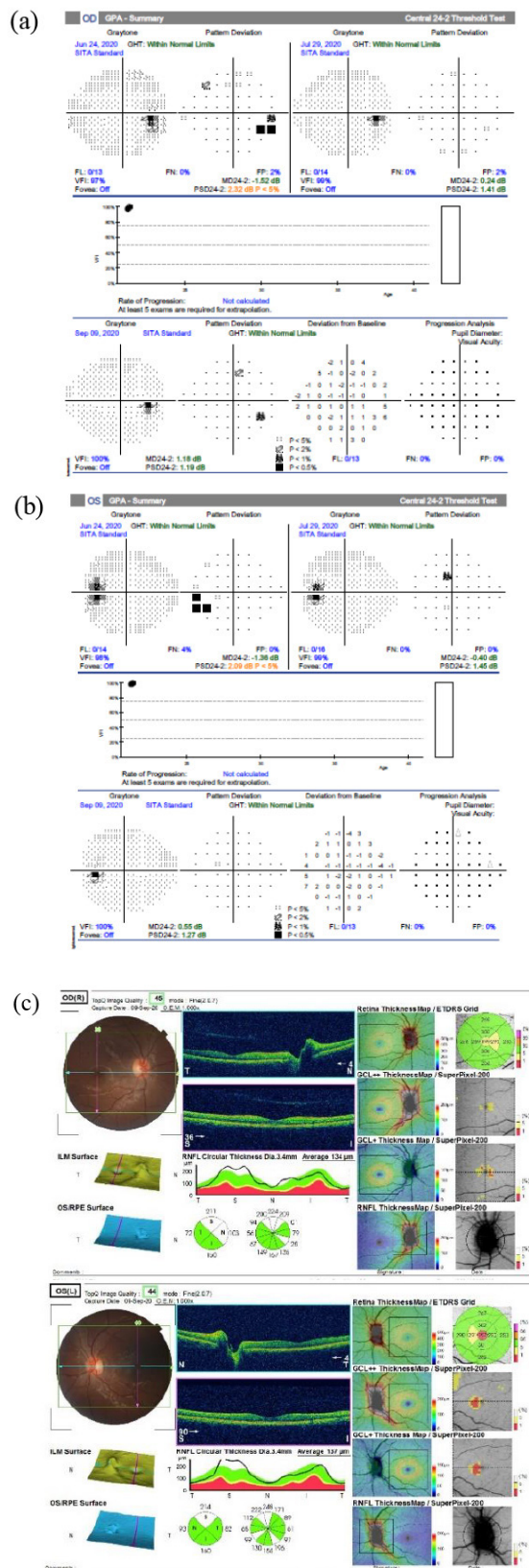


Figure 4. The result of the visual field from June to September with the Humphrey 24-2 shows an improvement on the right eye (a) and left eye (b), with no bilateral papilledema (c)

weight was 107 kg. There was no enlarged blind spot with Humphrey 24-2 visual field test and reduced papilledema with OCT (Figure 2). The results of MRI of the brain and orbits with contrast shown no abnormalities (Figure 3). The patient was consulted to the neurology department and the result was a patient treated with steroid oral and no needed for surgery. The patient was diagnosed with IIH with improvement in both eyes. Patient treated with acetazolamide, neurotropic and neuroprotective agents.

In August 2020, the patient returned for follow up with VA 6/6 RLE. IOP 12 mmHg RLE, and weight 103 kg. There was no enlarged blind spot with Humphrey 24-2 visual field test, and the OCT was good. The patient is still treated with acetazolamide, neurotropic and neuroprotective agents.

The patient returned to follow up again on September 2020 with improved visual field complaints and no headaches. The patient felt happy because the complaints were reduced, and her weight was 99 kg. VA was 6/6 RLE and IOP 18 mmHg RE and 14 mmHg LE. The Humphrey 24-2 visual field test and OCT were significantly improved compared to the patient first arrived (Figure 4).

DISCUSSION

Most of IIH patients come with severe headache, visual disturbances and bilateral papilledema.^{7,10,11} Headache is around 93% of patients with IIH.¹² Headache causes of IIH, as described by the International Classification of Headache Disorders (ICHD), 3rd edition (Figure 5). Reported from the Birmingham group, the headaches also determine the impaired quality of life.¹³

Papilledema patients who experience visual obscuration can manifest to increase in ICP, retinal ischemia or transient ischemia at the optic nerve caused by papilledema.¹⁴ Dizziness, nausea, diminished memory and concentration with horizontal diplopia due to sixth nerve palsy were other symptoms and clinical signs.^{7,10,11} Recent studies demonstrated a marked cognitive dysfunction and indicated that impaired executive function, memory, processing speed and reaction time remained after normalization of ICP and alleviated the headache and also the visual symptoms.^{7,15}

Funduscopy is the neuro-ophthalmological investigation of a suspected IIH patient. Test of vision and visual field by perimetry is also important, but the test method should be standardized.^{9,11,16} OCT was recommended for papilledema quantification and additional methods for papilledema detection

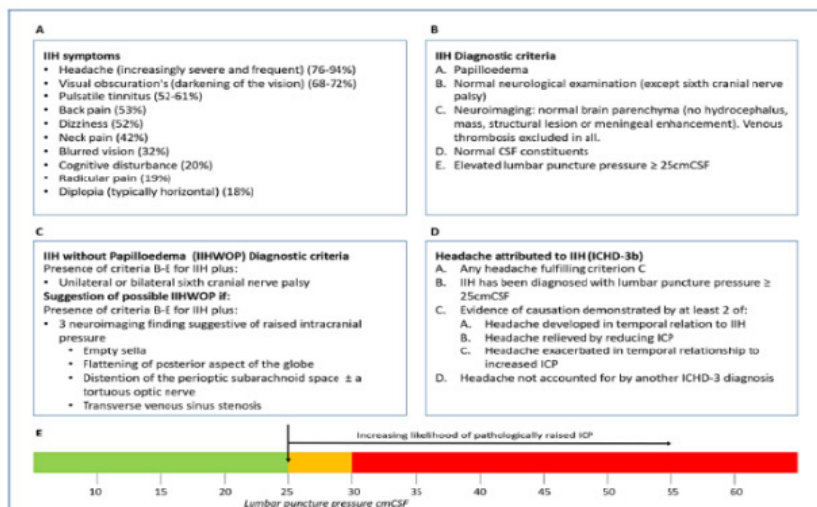


Figure 5. The consensus in diagnosing IIH. IIH diagnostic criteria and idiopathic intracranial hypertension without papilloedema (IIHWOP) diagnostic criteria.²²

and monitoring.^{16,17} RNFL measurements from time-domain OCT were found to correlate well with the clinical examination and the severity of visual dysfunction.^{18,19} Macular structure imaging containing the retinal ganglion cell layer (RGCL) and inner plexiform layer (IPL) has demonstrated little change over time, although optic nerve edema changes intracranial pressure. It is thought that the RGCL-IPL complex may more accurately represent long-term visual function and may potentially act as a structural biomarker of neuronal tissue loss. While further longitudinal studies are required to fully understand potential applications, existing OCT modalities and volumetric analyses have proven useful in providing a quantitative model of assessing the effects of IIH therapy.^{20,21}

Neuroimaging, preferably a brain MRI scan. MR/CT venography is required to rule out sinus venous occlusions. It remains uncertain if bilateral transverse sinus stenosis plays a role in IIH pathogenesis or in the radiological marker of elevated ICP. Empty sella, flattening of the posterior part of the globe and frequent subarachnoid space enlargement were the other neuroimaging anomalies of IIH.²²

In IIH management, weight loss has played a major role. This was based on case reports, but was recently confirmed in a prospective cohort study.²³ Twenty-five IIH patients were treated for three months with a deficient calorie diet (425 kcal/day). Patients decreased their weight significantly (15% of body weight) after dieting, resulting in a significant reduction in ICP with headaches and papilloedema. In another study, relative to patients without weight

loss, CSF pressure decreased markedly in patients with a BMI reduction of about 3.5 percent. The average weight loss was impressive with a 45.4 kg decrease, and the BMI was reduced by 16 kg/m².²⁴

Acetazolamide, an old diuretic with a substantial carbonic anhydrase inhibitor effect, is the first drug for IIH management. Two randomized trials have studied acetazolamide in IIH. The first randomized pilot study by Ball and colleagues on acetazolamide versus no acetazolamide.²⁵ The second research was a multicenter, randomized, double-masked, placebo-controlled study of acetazolamide in 165 IIH participants and found that acetazolamide had a small but important beneficial impact on visual function.²⁴ Furosemide and other diuretics, either alone or with acetazolamide, are often used in IIH, but no randomized controlled trials confirm its effectiveness. As a management choice in IIH, Topiramate has become increasingly popular. The combined effect as carbonic anhydrase inhibitor, although weaker than acetazolamide, was the preventive on migraine and side effect of appetite suppression and weight loss. In addition to dizziness, balance, and gait issues, the recorded adverse effects are very similar to acetazolamide, with paresthesia, weakness, and gastrointestinal symptoms. In a prospective acetazolamide study, The prevalence of headache was reduced from 68% to 43% in acetazolamide-treated patients, compared to a reduction from 72% to 65% in the control group.²⁵ However, no significant treatment effects were noted. In the prospective follow-up study by Yri and colleagues, 43% of patients either recovered completely or reported only infrequent headaches (≤ 1 day/month).⁷

IIH appears to be a complex illness. The multiple medical specialties required to be involved, and the relapse rate is high (up to 40%).²⁵ Visual function was almost normalized in the IIH population in all subjects after 12 months, while headache and diminished memory were the main long-term complications.⁷ Therefore, patients education and near long-term follow-up with weight management are recommended.

CONCLUSION

IIH is a challenging and severe disease that require a collaboration of department s consisting of a neuro-ophthalmologist, neurologist and nutritionist, and follow-up visits to patients. In IIH patients, the visual prognosis is usually good, the possibility of permanent visual loss exists and remains a significant cause of this disease process's morbidity. Early and aggressive management is required to prevent irreversible visual impairment.

CONFLICT OF INTEREST

The authors declare no competing financial interest.

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None

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