Original Article

Benign Idiopathic Intracranial Hypertension with Empty Sella and Gonadotrophin Insufficiency

Nida Hafeez[1], Nayab Gull[2], Ambreen Gull[3], Fuad Ahmad Khan Niazi[4]

Abstract

Background: Benign idiopathic intracranial hypertension (BIIH) is characterised by increased intracranial pressure without identifiable intracranial pathology and commonly presents with headache. However, atypical presentations without headache may delay the diagnosis and increase the risk of irreversible visual loss. Empty sella syndrome is a radiological finding that may be associated with increased intracranial pressure and pituitary dysfunction, although hormonal abnormalities are often under-recognized.

Case Presentation: We report the case of a 38-year-old woman who was obese and presented with progressive visual impairment and asymmetric bilateral papilledema in the absence of headache, nausea, or vomiting. Visual acuity was preserved in the right eye but markedly reduced in the left eye. Neuroimaging revealed an empty sella with flattening of the pituitary gland, and cerebrospinal fluid opening pressure was markedly elevated. Hormonal evaluation demonstrated isolated gonadotropin insufficiency with low follicle-stimulating hormone levels. The patient was diagnosed with benign idiopathic intracranial hypertension associated with empty sella syndrome. Management included therapeutic lumbar puncture, oral acetazolamide, and combined oestrogen–progesterone hormone replacement therapy, which was initiated after an endocrinology consultation. Gradual improvement in visual function was observed over a six-month follow-up period.

Conclusion: This case highlights an atypical, headache-free presentation of benign idiopathic intracranial hypertension associated with empty sella syndrome and gonadotropin insufficiency. It underscores the importance of considering BIIH in patients presenting primarily with visual symptoms and emphasises the role of comprehensive hormonal evaluation and multidisciplinary management in optimising visual outcomes.

Keywords: Idiopathic Intracranial Hypertension; Empty Sella Syndrome; Papilledema; Gonadotropin Deficiency; Visual Impairment.

Introduction

Benign idiopathic intracranial hypertension (BIIH) is a disorder characterised by elevated intracranial pressure in the absence of an identifiable intracranial mass, hydrocephalus, or central nervous system infection. It predominantly affects overweight women of reproductive age and classically presents with headache, transient visual obscurations, pulsatile tinnitus, and papilledema.[1] Visual impairment is the most serious complication of BIIH and may become irreversible if the diagnosis and treatment are delayed. Although headache is the most common presenting symptom, atypical presentations without headache have been increasingly recognised and may pose a diagnostic challenge, particularly when visual symptoms predominate.[2]

Empty sella syndrome (ESS) is a radiological condition resulting from herniation of the subarachnoid space into the sella turcica, leading to partial or complete flattening of the pituitary gland. Chronic elevation of intracranial pressure is believed to play a key role in the development of ESS by exerting downward pressure on the diaphragma sellae and pituitary gland. Consequently, ESS has been frequently reported in association with BIIH, suggesting a shared pathophysiological mechanism rather than a coincidental finding.[3]

Although many patients with ESS remain endocrinologically asymptomatic, pituitary dysfunction may occur due to compression of the pituitary tissue, most commonly affecting the gonadotrophic axis. Hormonal abnormalities in patients with BIIH and ESS are often under-recognized, particularly in the absence of overt clinical symptoms, such as menstrual irregularities or infertility. Consequently, isolated gonadotropin insufficiency may remain undetected unless a comprehensive hormonal evaluation is conducted.[4]

We report this case to highlight an atypical presentation of BIIH in a patient who presented without headache but with progressive visual impairment and asymmetric papilledema in association with empty sella syndrome and isolated gonadotropin insufficiency.[5,6] The absence of documented menstrual complaints, coupled with significant visual involvement, underscores the importance of considering endocrine evaluation in patients with BIIH and ESS. This case further demonstrates the role of multidisciplinary management in achieving visual improvement and contributes to the limited literature describing hormonal dysfunction in patients with BIIH-associated empty sella syndrome.

Case Presentation

A 38-year-old woman, who was obese (body mass index, 32), presented with a gradual progressive decline in vision for approximately six months. She primarily complained of blurred vision and visual field disturbance, which was more pronounced in the left eye. Notably, she denied any history of headache, nausea, vomiting, or transient visual obscurations. There was no prior history of ocular disease, ocular trauma, or significant family

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history of neurological or ophthalmic disorders. She was married and had two children, with no documented menstrual irregularities.

On ophthalmic examination, the best-corrected visual acuity was 6/6 in the right eye and counting fingers at one foot in the left eye. Fundus examination revealed bilateral papilledema with marked asymmetry; the left optic disc showed pallor with severe swelling, whereas the right optic disc demonstrated mild disc oedema. Automated perimetry revealed bilateral enlargement of blind spots with associated nasal visual field loss.

Initial neuroimaging with a non-contrast computed tomography (CT) scan of the brain was performed to exclude intracranial mass lesions and was unremarkable. Subsequent non-contrast magnetic resonance imaging (MRI) of the brain and orbits demonstrated herniation of the subarachnoid space into the sella turcica with flattening of the pituitary gland, consistent with an empty sella. Given these findings, a hormonal evaluation was undertaken, which revealed reduced follicle-stimulating hormone levels consistent with isolated gonadotropin insufficiency, while other pituitary hormone levels were within normal limits.

A diagnostic lumbar puncture revealed an elevated cerebrospinal fluid opening pressure of 40 cm H_2O with normal cerebrospinal fluid composition. Based on the clinical presentation, radiological findings, and elevated intracranial pressure, a diagnosis of benign idiopathic intracranial hypertension associated with empty sella syndrome and gonadotropin insufficiency was established. Written informed consent was obtained from the patient for the publication of this case report and the accompanying images.

The patient was treated with a single therapeutic lumbar puncture and commenced on oral acetazolamide at a dose of 250 mg four times daily. She was referred to the Endocrinology Department, where combined hormone replacement therapy was initiated in the form of oral oestradiol valerate 2 mg daily along with medroxyprogesterone acetate 10 mg daily for 10 days each month. Lifestyle modification, including dietary counselling and advice regarding weight reduction, was also provided.

Results

At presentation, the best-corrected visual acuity was 6/6 in the right eye and counting fingers at one foot in the left eye. Fundus examination demonstrated bilateral papilledema with marked asymmetry, which was more severe in the left eye. Automated perimetry showed bilateral enlargement of the blind spots with associated nasal visual field defects.

Over the six-month follow-up period, visual acuity in the left eye improved from counting fingers at one foot to 6/60, whereas visual acuity in the right eye remained stable at 6/6. Serial fundoscopic examinations demonstrated complete resolution of optic disc oedema in the right eye, while the left eye showed resolution of disc swelling with residual optic disc pallor. Repeat VF testing was not performed; however, the patient reported subjective improvement in visual perception corresponding with clinical improvement in optic disc appearance.

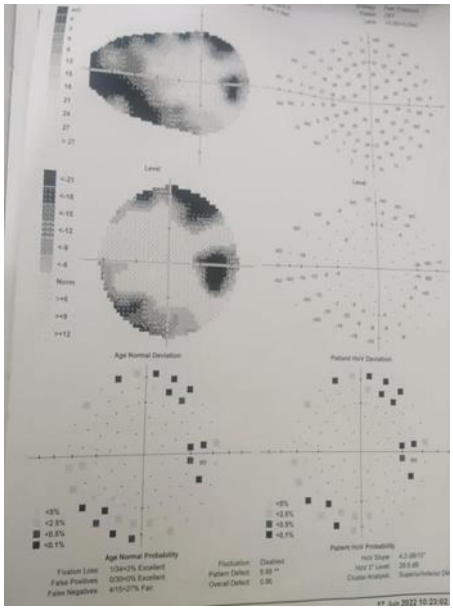

Figure 1: Visual Field Test of Right Eye

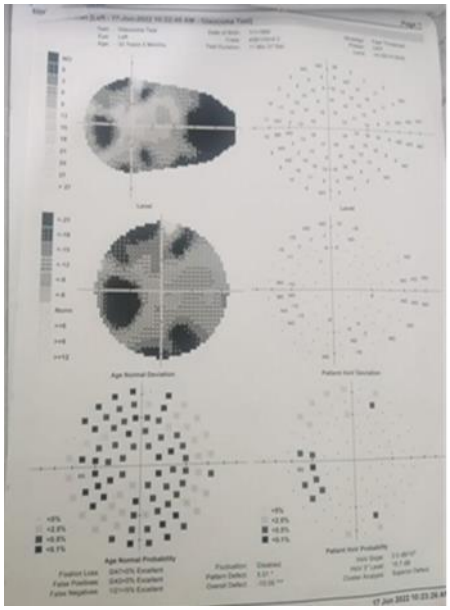

Figure 2: Visual Field Test of Left Eye

Discussion

Benign idiopathic intracranial hypertension is an important cause of potentially reversible visual loss, particularly in overweight women of reproductive age. Persistent elevation of intracranial pressure can result in optic nerve head oedema and progressive optic neuropathy if not promptly recognised and treated.[9] Although headache is the most frequently reported symptom of BIIH, a subset of patients may present without headache, as seen in this case, leading to a delayed diagnosis and increased risk of visual morbidity. The presence of asymmetric papilledema and marked unilateral visual impairment further contributed to the diagnostic complexity in this patient.[5]

The association between BIIH and empty sella syndrome has been increasingly recognised in the literature. Empty sella is believed to result from chronic elevation of intracranial pressure, causing herniation of the subarachnoid space into the sella turcica and subsequent compression and flattening of the pituitary gland. Several imaging studies have demonstrated a high prevalence of empty sella in patients with BIIH, supporting a

shared pathophysiological mechanism rather than coincidental coexistence. However, the clinical implications of this association, particularly with respect to pituitary function, are often overlooked.[7]

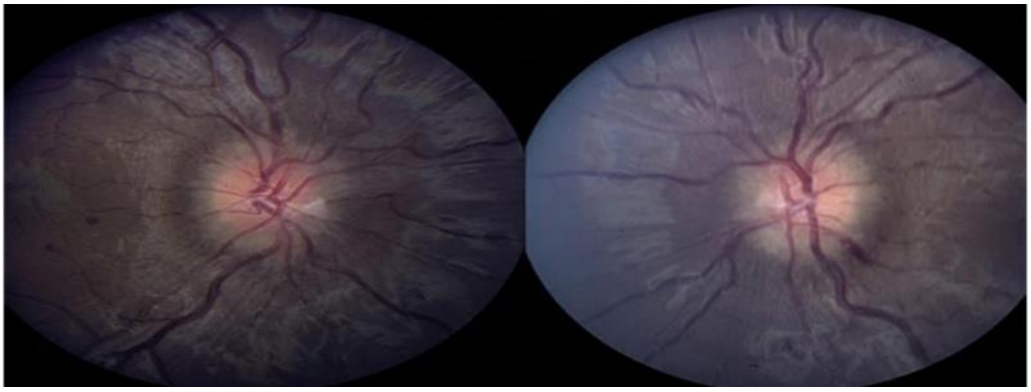

Figure 3: Fundus Photograph of Right and Left Eye

Most patients with empty sella syndrome remain endocrinologically asymptomatic, and when hormonal abnormalities are present, they typically involve multiple pituitary axes. Isolated gonadotrophin insufficiency, as observed in this patient, is uncommon and may remain undetected in the absence of overt clinical features, such as menstrual irregularities or infertility.[8] The absence of documented menstrual complaints in this case emphasises the need for proactive hormonal assessment in patients with BIIH and radiological evidence of an empty sella, even when classic endocrine symptoms are lacking.[2]

MRI BRAIN&ORBIT

Clinical Info:

Empty sella is noted.

IMPRESSION:

- Empty sella otherwise essentially unremarkable pre and post-contrast MR study of brain and orbits.

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Figure 4: MRI Brain + Orbit (Report)

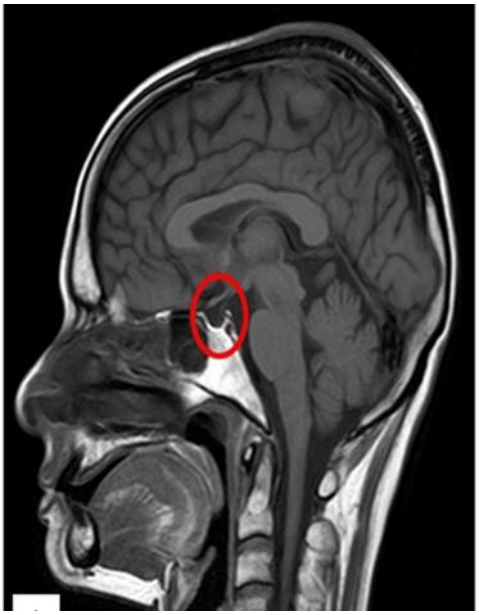

Figure 5: MRI Brain + Orbit Showing Empty Sella

A limited number of reported cases in the literature describe the coexistence of BIIH, empty sella syndrome, and pituitary hormonal dysfunction. Management strategies in these cases have focused on reducing intracranial pressure through medical therapy with acetazolamide, therapeutic lumbar puncture, weight reduction, and, in refractory cases, cerebrospinal fluid diversion procedures.[2] Visual improvement has been reported over variable timeframes ranging from weeks to several months. In the present case, a multidisciplinary approach involving ophthalmology, neurology, and endocrinology resulted in quantifiable visual recovery over six months following treatment with acetazolamide, therapeutic lumbar puncture, and combined oestrogen–progesterone hormone replacement therapy.[10] This case underscores the importance of maintaining a high index of suspicion for benign idiopathic intracranial hypertension in patients presenting with isolated visual symptoms, highlights the need for routine endocrine evaluation in the presence of empty sella syndrome, and reinforces the value of a multidisciplinary approach in preventing irreversible visual loss.[12]

Conclusions

Benign idiopathic intracranial hypertension may present without headache and predominantly with visual symptoms, increasing the risk of delayed diagnosis and irreversible visual loss. The coexistence of empty sella syndrome and isolated gonadotropin insufficiency highlights the

importance of comprehensive endocrine evaluation in patients with BIIH. Early recognition and a multidisciplinary management approach involving ophthalmology, neurology, and endocrinology can lead to meaningful visual recovery and improved overall outcomes.[11]

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