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Peculiar case of idiopathic intracranial hypertension. Case report

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Abstract

Background

Idiopathic Intracranial Hypertension (IIH) is a neurological disorder characterized by elevated intracranial pressure without any detectable intracranial mass or hydrocephalus. It typically presents in women of childbearing age and may manifest with symptoms like headache, visual disturbances, and papilledema. While the aetiology remains unknown, several systemic factors such as obesity, endocrine abnormalities, renal dysfunction, and hypertension have been associated with worsening of the clinical presentation and complexity in management.

Case presentation

A 36-year-old female with a known history of hypertension presented with persistent headache and visual complaints. On clinical examination, her blood pressure was markedly elevated at 220/110 mmHg. Laboratory evaluation showed deranged renal function with blood urea at 85 mg/dL and serum creatinine at 4.8 mg/dL. Ophthalmic examination revealed bilateral papilledema. Neuroimaging was promptly performed to rule out intracranial mass lesions or venous thrombosis, which confirmed the diagnosis of IIH.

Multidisciplinary input from nephrology, neurology, and ophthalmology was essential to manage the complex interplay between intracranial hypertension, hypertension, and renal compromise.

Conclusion

This case highlights an uncommon presentation of IIH in a patient with coexisting renal dysfunction and severe hypertension. While IIH is generally idiopathic, systemic factors can contribute significantly to its manifestation and prognosis. Renal dysfunction, in particular, not only complicates the use of first-line therapies like carbonic anhydrase inhibitors but also may be independently associated with altered cerebrospinal fluid dynamics and increased intracranial pressure.

Keywords: *Idiopathic Intracranial Hypertension, papilledema, renal dysfunction*
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Introduction

This is a case report of Idiopathic intracranial hypertension(IIH), formerly known as pseudotumor cerebri, a condition characterized by increased intracranial pressure (ICP) without an identifiable cause on neuroimaging or cerebrospinal fluid analysis, which is a known entity among females, especially in the reproductive age.[1] Over the years, numerous studies, like the Idiopathic Intracranial Hypertension Treatment Trial (IIHTT) conducted by Wall et al., have expanded the understanding of IIH with respect to epidemiology, diagnostic markers, and management strategies.[1] While obesity remains the strongest risk factor, some studies have also suggested the associations with endocrine abnormalities, vitamin A toxicity, and oral contraceptives. Chronic Kidney disease remains a rare cause or a risk factor [2, 3]. This case is one of its kind, where, despite having a diagnosis of IIH, the management of the patient posed a challenge for the clinicians.

Case Report

A 36-year-old female presented to a nephrologist with complaints of headache 15 days ago. Her blood pressure

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(BP) was recorded as 220/110 mmHg at the time of presentation. Laboratory investigations showed a deranged value of hemoglobin 9.3mg/dl, serum urea 86, serum creatinine 4.6, so she was admitted to the nephrology department for further evaluation. The patient had also been a case of stress-induced gestational hypertension four years ago, with urea and creatinine values being 46 and 1.4, respectively. Patient had been managed successfully with a conservative approach during her pregnancy, with her antenatal, natal, and post-natal period being uneventful. The rise in BP, mostly induced by stress, was noted by the patient three years ago, for which she would take antihypertensives on and off, but not regularly. This time, her concern for visiting the hospital was a deranged value of urea and creatinine.

The patient was sent to the eye department for ocular assessment and fundus evaluation, as the patient complained of blurred vision in the right eye. Patient was conscious, cooperative, and well oriented to time, place, and person. Her visual acuity was RE: 6/9 (with -0.25×180 -6/6); Left eye (LE): 6/9 (with -0.50×180 -6/6) using Snellen's chart. Intraocular Pressure (IOP) by Non Contact Tonometry (NCT) was recorded to be 12 mmHg in both eyes with a normal pupillary reaction and no RAPD in either eye. Patient was willing to perform color vision, which was found to be normal in both eyes (BE). Visual fields of both eyes were repeated multiple times to get accurate results, and it was found that the Visual fields in both eyes were constricted, more in the right eye than the left eye. A dilated fundus examination revealed bilateral disc edema, no hemorrhages or hard exudate, and no evidence of venous stasis or papilledema. On orbital sonography, the optic nerve acoustic shadow of both nerves was widened, with the optic nerve of RE being more than that of LE. Patient was diagnosed as Papilledema BE (Frisen scale Grade 3 RE and Grade 1 LE, respectively). [Figure 1] The possibility of Hypertensive Retinopathy was ruled out due to the absence of exudates, hemorrhages, and arteriovenous changes. Brightness Scan ultrasonography of both the eyes shows enlargement of the optic nerve head (right eye >left eye), suggesting papilledema. [Figure 2] Her MRI scan had been suggestive of "Partially Empty Sella", [Figure 3] and MRV ruled out venous sinus thrombosis, so the papilloedema in her case was attributed to Idiopathic Intracranial Hypertension. A fundus photograph was taken on a TRC-50DX fundus camera for documentation and comparisons in the future.

She was already being managed on an antihypertensive drug (tablet telmisartan 5 mg once daily) by the nephrology department, which had no adverse effects on the patient. Through the treatment, her blood pressure (BP) had lowered, and her urea/creatinine was found to be 63/4.2, after nearly one month of observation. Lumbar Puncture investigation for Cerebrospinal Fluid (CSF) analysis was not done.

Repetitive IOP readings remained 12mmHg in both eyes, while visual acuity improved by a line (6/6) and clarity in both eyes during her stay at the hospital. Fundus examination revealed papilledema BE (Frisen scale Grade 3 RE and Grade 1 LE, respectively) with no signs of disc hemorrhages, exudates, and arteriovenous changes. A repeat MRV done after a month of observation revealed no significant abnormality. The patient is kept under observation for vision, color vision, intraocular pressure, visual fields, and fundus examination.

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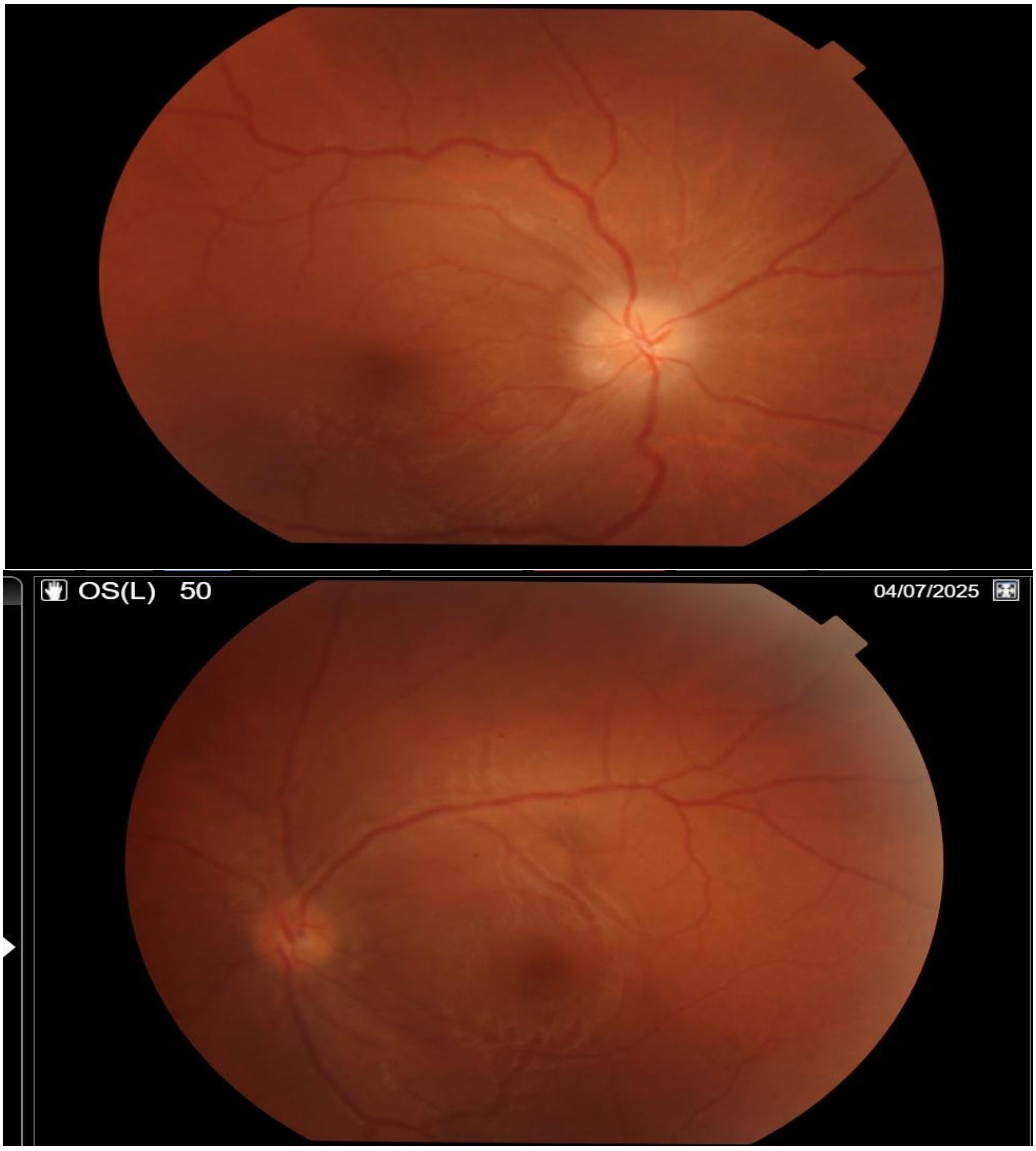

Figure 1: Grade 3 papilledema with disc elevation and obliteration of blood vessels (right eye)

Grade 2 papilledema with disc elevation and obliteration of blood vessels (left eye)

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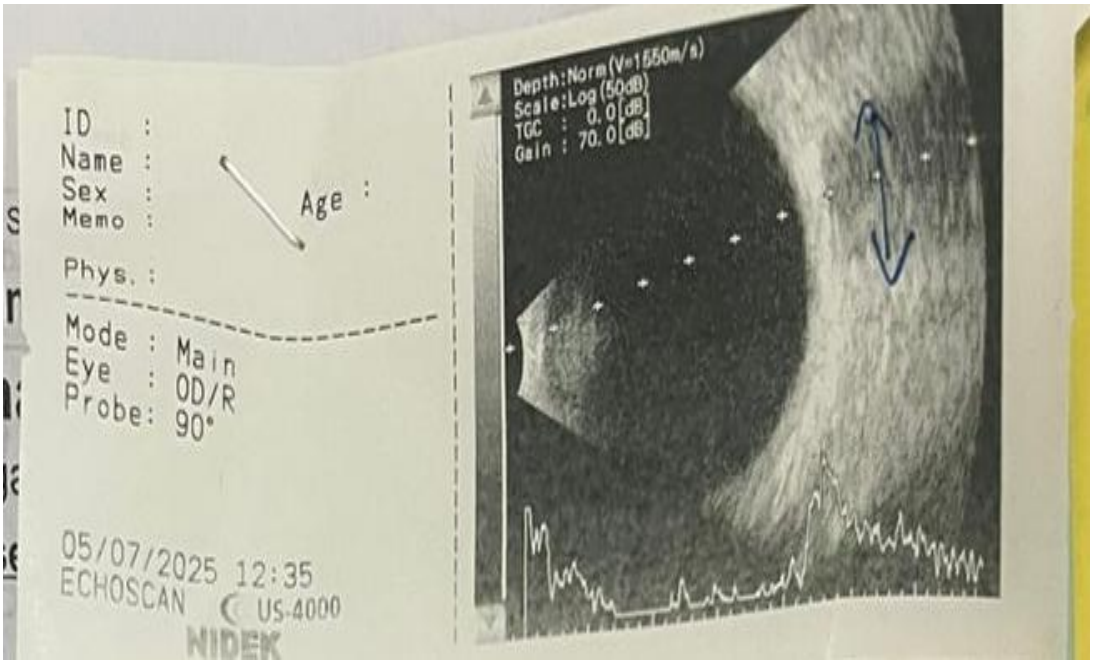

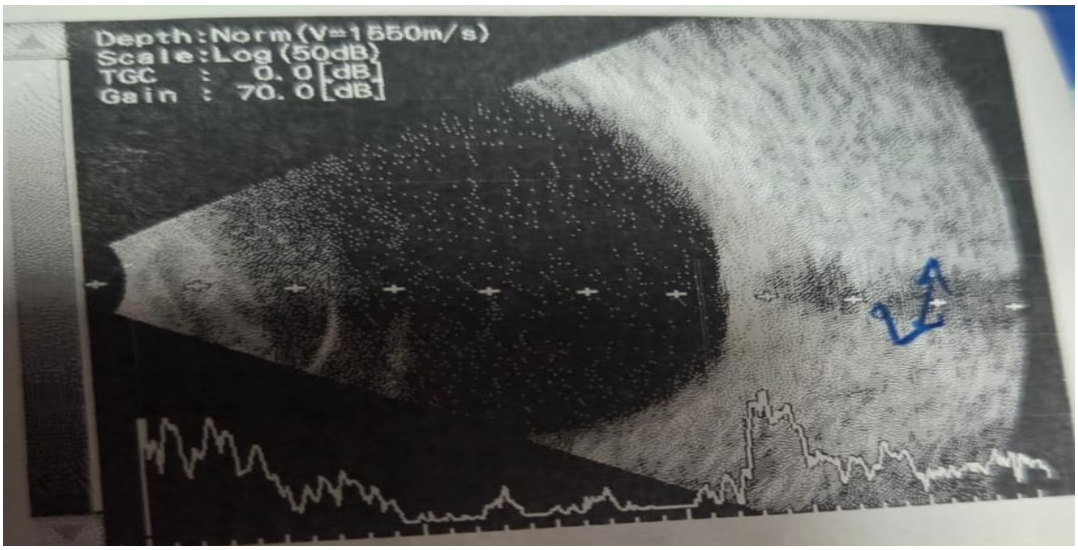

Figure 2: B scan suggestive of enlarged optic nerve head (right eye > left eye)

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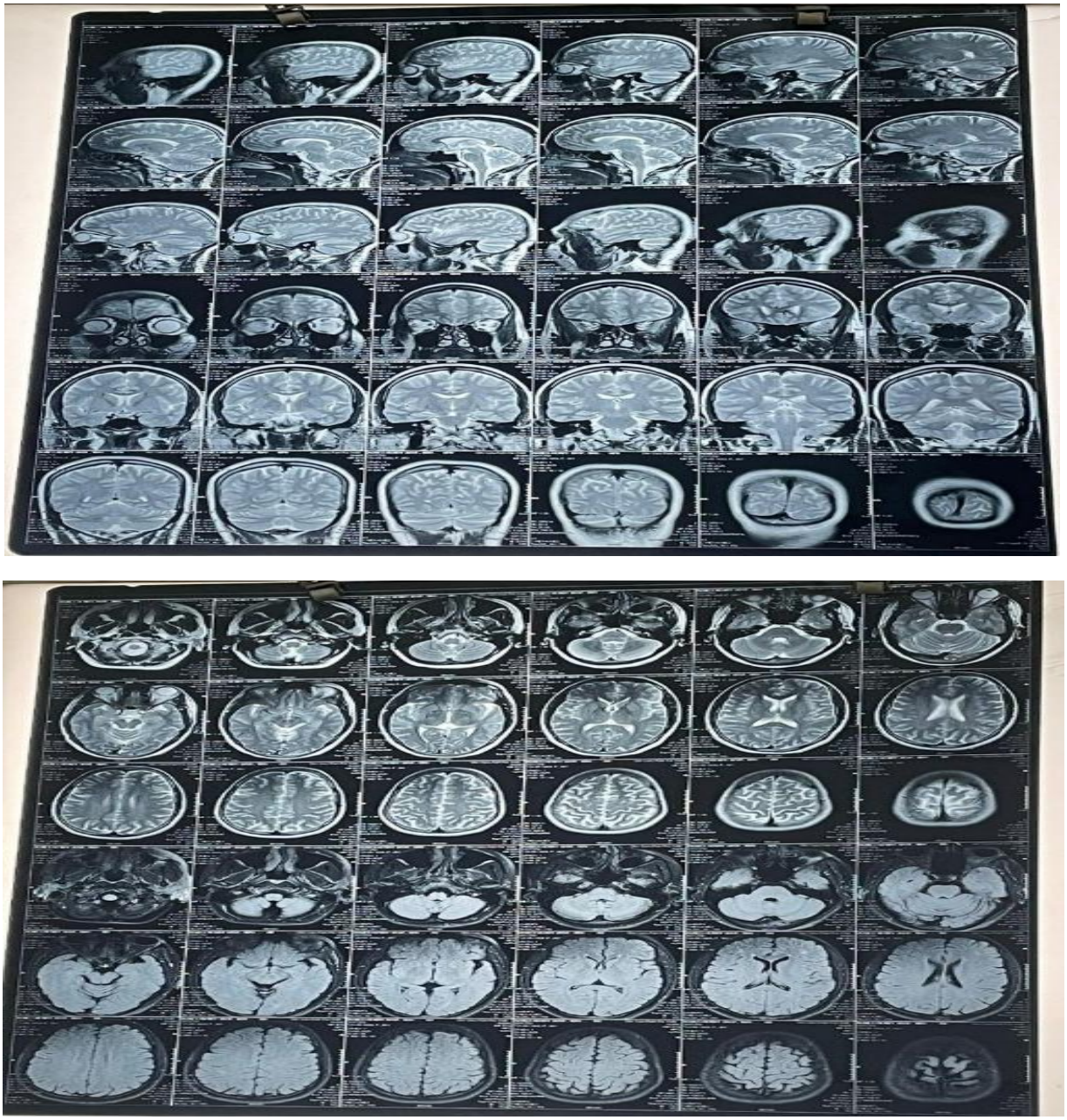

Figure 3: MRI scan had been suggestive of "Partially Empty Sella"

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Discussion

Imaging features such as empty sella in 34.4% of cases, dilated optic sheath in 12.5%, tortuous optic nerve in 18.8%, posterior flattening of sclera in 9.4% and all of the above in 3.1%, have been frequently reported in IIH patients.[4,5,6] A study by Yuh et al., conducted in the Medical Research Council (UK), identified partially empty sella in 70% of IIH patients, a feature also present in the current case.[7] Geeraerts et al. in their study have also demonstrated the utility of bedside optic nerve sheath diameter (ONSD) ultrasonography as a non-invasive marker of raised ICP, with good correlation to invasive monitoring techniques. [8] In this case, it can be well noticed in her ocular sonography pictures. Our patient showed a constriction of visual fields in the initial stages, as mentioned in several studies. In a study by Ambika et al., normal visual field was seen in 12% of cases, nasal and arcuate defects in 7% and advanced generalized constricted fields in 11%.[9] In a study by Claire Chagot et al., visual field defects were found in 50% cases.[10]

The patient was lean, and due to her perpetually raised urea and creatinine levels, Acetazolamide, as a drug, could not be considered as an optimum choice. Patient has been managed conservatively, with fluids and monitoring.

Conclusion

This has been a rare case where a young female patient had a deranged value of urea and creatinine despite optimum management of Hypertension. "Stress" has emerged as a trigger for a rise in BP, but this case shows the deep impact that small episodes of stress could have on young patients, even after adequate control of hypertension. With such sensitive findings in the optic disc, combined with refractory parameters of urea and creatinine, management of IIH becomes a challenge.

Takeaway message

This case highlights the rare coexistence of idiopathic intracranial hypertension (IIH) with renal dysfunction in a young, non-obese female, where stress-induced hypertension further complicated management. It underscores the diagnostic value of multimodal imaging and ocular evaluation, and the therapeutic challenge when first-line agents like acetazolamide are contraindicated. Individualized, conservative monitoring remains crucial in such complex presentations.

conflict of interest

No conflict of interest

Source of funding

No funding taken

Ethical approval

Ethical approval was not required in the above case.

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No availability of data and material

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