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Case Series

Atypical Presentations of Idiopathic Intracranial Hypertension in Clinical Practice. A Case Series

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Abstract

Idiopathic intracranial hypertension (IIH) usually presents with headache, papilledema, and sixth nerve palsy, but atypical manifestations can delay diagnosis. Atypical presentations in the present case series were trigeminal neuralgia, focal seizures with Todd's paresis, hypoglossal nerve palsy, intractable migraine like headache, orthostatic intolerance and spontaneous CSF rhinorrhea. Most patients improved with medical therapy, with only two requiring surgical intervention. These findings emphasize that atypical IIH is not uncommon and that measuring cerebrospinal fluid opening pressure is essential for timely diagnosis.

Keywords: *Idiopathic intracranial hypertension, Atypical neurological manifestations, Cerebrospinal fluid pressure, Papilledema*

Introduction

Idiopathic intracranial hypertension (IIH) is characterized by raised intracranial pressure (ICP) without an identifiable structural cause. Classic clinical features include headache, papilledema, and diplopia from sixth nerve palsy, transient visual obscurations, and pulsatile tinnitus. [1] Atypical presentations, though recognized in literature, remain underdiagnosed and may lead to prolonged diagnostic pathways.

Reported atypical manifestations include IIH without papilledema (IIHWOP), unilateral or highly asymmetric papilledema, trigeminal neuralgia, facial palsy, olfactory dysfunction, auditory and vestibular symptoms, seizures, radiculopathy, and spontaneous skull base CSF leaks. [2] Other very rare atypical presentation of IIH is pseudo foster Kennedy syndrome [3] and cyclic vomiting syndrome [4] such presentations warrant consideration of IIH, particularly in young obese women, even in the absence of classic signs. There are few case reports of atypical presentation of IIH from India [3, 5, 6], Saudi Arabia [7, 8], USA [4] and Germany [9].

Case Descriptions

Case 1: Trigeminal Neuralgia as a Manifestation of IIH

A 45-year-old woman presented with chronic right-sided facial pain for five years, localized to the maxillary (V2) distribution of the trigeminal nerve. Her pain was sharp, episodic, and triggered by touch, consistent with trigeminal neuralgia. Neurological examination was otherwise normal, with no papilledema. MRI brain with venography revealed a partial empty sella and kinking of the optic nerves. CSF opening pressure (OP) was 27 cm H_2O, confirming elevated intracranial pressure. A diagnosis of atypical IIH presenting with trigeminal neuralgia was made. The patient responded well to acetazolamide therapy.

Case 2: Focal Motor Seizures with Todd's Paresis

A 48-year-old woman with longstanding hypertension and hypothyroidism presented with recurrent right brachial focal motor seizures, occasionally followed by Todd's paresis. Symptoms had persisted for 12 months. Neuroimaging demonstrated a complete empty sella, enlarged optic nerve sheath, and a prominent Meckel's cave, suggesting chronically elevated intracranial pressure. CSF OP measured 25 cm H_2O. There was no papilledema. The patient was diagnosed with IIH presenting atypically with seizure activity. She improved on acetazolamide and topiramate.

Case 3: IIH without Papilledema (IIHWOP presenting as chronic headache

A 39-year-old woman with hypothyroidism and vitamin D deficiency reported chronic headache for 14 months. Fundus examination showed no papilledema. MRI brain showed partial empty sella, mild tortuosity of both optic nerves, and increased perioptic CSF spaces. CSF OP was 27 cm H_2O, supporting the diagnosis of IIHWOP. The patient demonstrated symptomatic improvement on acetazolamide therapy.

Case 4: CSF Rhinorrhea Revealing Underlying IIH

A 40-year-old woman developed clear watery nasal discharge immediately after a nasopharyngeal COVID-19 swab test. Beta-2 transferrin testing confirmed CSF rhinorrhea. CT cisternography showed a 1.7-mm skull base defect in the left posterior cribriform plate. MRI revealed empty sella, increased perioptic CSF spaces, optic nerve kinking, and bilateral transverse sinus stenosis. CSF OP was significantly elevated at 34 cm H_2O. The patient underwent surgical repair of the skull-base defect and was subsequently managed for IIH with acetazolamide.

Case 5: Hypoglossal Nerve Palsy with Dysarthria

A 47-year-old woman with hypertension presented with acute dysarthria and left-sided headache of 7 days' duration. Examination revealed left hypoglossal (XII) nerve palsy: the tongue deviated to the left on protrusion and to the right at rest, consistent with lower motor neuron involvement. MRI brain showed complete empty sella and optic nerve sheath enlargement. CSF OP was 28 cm H_2O. This rare example of IIH presenting with isolated hypoglossal palsy responded to acetazolamide and blood pressure control.

Case 6: Orthostatic Intolerance Associated with IIH

A 24-year-old woman with hypothyroidism presented with intermittent, brief giddiness on standing, occurring over 2 months. She denied significant headache or visual symptoms. MRI revealed complete empty sella and optic nerve sheath distension. Her CSF OP was markedly elevated at 39 cm H_2O. Despite minimal headache, imaging and CSF findings supported the diagnosis of IIH. The patient improved on acetazolamide with emphasis on volume status and postural precautions.

Case 7: Possible IIHWOP with Migraine-Like Headache

A 19-year-old morbidly obese woman presented with migraine-like headaches without aura for two years. She had no papilledema. MRI demonstrated complete empty sella and optic nerve sheath enlargement. CSF OP was 44 cm H_2O, confirming significantly raised intracranial pressure. She was diagnosed with IIHWOP and treated with acetazolamide along with structured weight-reduction therapy. Since the headache was refractory a lumboperitoneal (LP) shunt was done.

Table 1. Showing summary of demographic data, duration of symptoms, atypical features and comorbidities in IIH cases.

Case	Age (yrs) / Sex	Duration of Symptoms	Atypical Presentation	Comorbidities
1	45 / F	5 years	Right trigeminal neuralgia (maxillary division)	Nil
2	48 / F	12 months	Focal motor brachial seizures with Todd's paresis	Hypertension, hypothyroidism
3	39 / F	14 months	IIH without papilledema (IIHWOP)	Hypothyroidism, vitamin D deficiency
4	40 / F	15 days	CSF rhinorrhea post–COVID nasal swab	Nil
5	47 / F	7 days	Dysarthria, left hypoglossal nerve palsy	Hypertension
6	24 / F	2 months	Orthostatic intolerance, episodic giddiness	Hypothyroidism
7	19 / F	2 years	Migraine-like headache (IIHWOP)	Morbid obesity

Table 2. Showing MRI findings and atypical features in IIH cases.

Case	MRI / MRV Findings	Atypical Presentation	CSF Opening Pressure (cm H_2O)
1	Partial empty sella, optic nerve kinking	Right trigeminal neuralgia (maxillary division)	Nil
2	Complete empty sella, enlarged optic nerve sheath, prominent Meckel's cave	Focal seizures	25
3	Partial empty sella, tortuous optic nerves, increased perioptic CSF	IIHWOP	27
4	Empty sella, increased perioptic CSF, ON kinking, b/l transverse sinus stenosis	Skull-base defect (1.7 mm), CSF rhinorrhea	34
5	Complete empty sella, optic nerve sheath distension	Hypoglossal palsy	28
6	Complete empty sella, increased optic nerve sheath	Orthostatic intolerance	39
7	Complete empty sella, optic nerve sheath enlargement	IIHWOP	IIHWOP

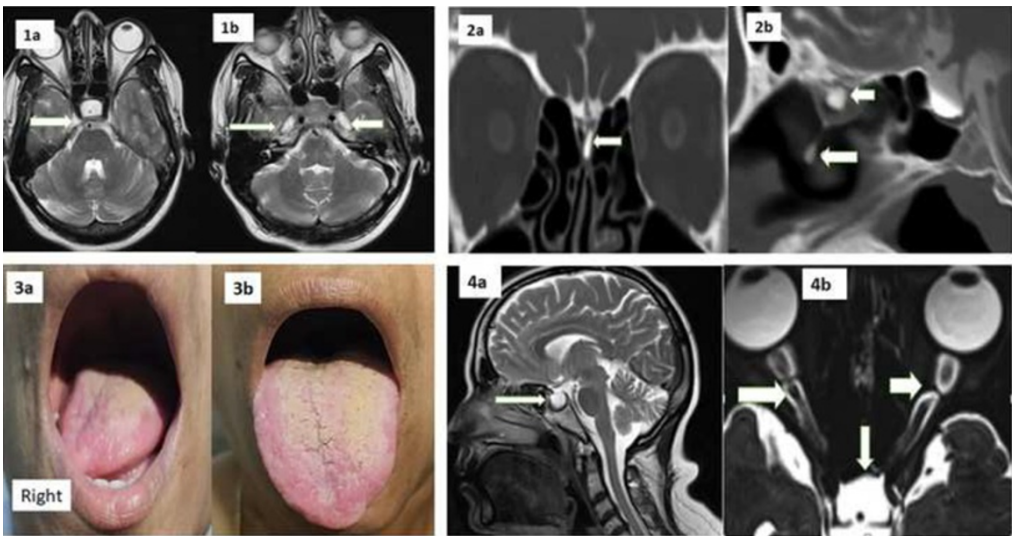

Figure 1. Magnetic Resonance Imaging (MRI) brain T2 axial section (1a,1b) showing bilateral Meckel's cave enlargement, Computed Tomography cisternography brain with contrast coronal (2a) and sagittal (2b) sections showing bony defects in the posterior cribriform plate with cerebrospinal fluid (CSF) leakage. Image showing deviation of tongue to right side in resting position (3a) and deviation to left side on protrusion of tongue (3b) s/o left 12th nerve palsy. MRI Image sagittal section showing complete empty sella (4a) and axial sections showing enlargement of perioptic sheath CSF space with tortuosity of optic nerve and flattening of posterior orbital globe in case of possible IIH without papilledema.

Treatment and Outcomes

Five patients were treated with acetazolamide. Two patients received a combination of acetazolamide and topiramate. Case 4 required transnasal surgical repair of a skull base defect and case 7 required LP shunt. Most patients experienced clinical improvement. No recurrences were noted during available follow-up.

Discussion

In this cohort, 12.9% (7) of IIH cases had atypical features, consistent with global literature indicating that atypical presentations comprise a substantial minority of IIH patients. A striking finding was that all affected individuals were women, reaffirming the well-established female predominance in IIH. [11]

The spectrum of atypical manifestations in this study aligns with reported literature, including trigeminal neuropathy, lower cranial nerve involvement, seizure disorders, and spontaneous CSF leaks. Hypoglossal palsy, as observed in Case 5, is particularly rare and highlights the potential of IIH to affect even lower cranial nerves. Similarly, enlargement of Meckel's cave in the seizure case echoes previous associations of chronically elevated ICP with bony remodeling. [6]

IIHWOP can pose a significant diagnostic challenge. Reliance on imaging markers and elevated CSF opening pressure is essential in such cases. Importantly, delayed diagnosis was common in this series, with symptom duration extending up to five years in one patient. Early recognition of atypical features could prevent prolonged morbidity and unnecessary diagnostic procedures.

The strong association between obesity and IIH was reaffirmed; weight reduction remains a cost-effective disease-modifying therapy. It was found that Indian cases of typical IIH were not obese [11] so presence of atypical presentation in our cases, obesity was an important association. To our knowledge the present case series is largest series of IIH with atypical presentation.

Conclusion

Atypical IIH can mimic other neurological disorders. Clinicians should suspect IIH in obese women presenting with unexplained cranial neuropathies, trigeminal neuralgia, CSF rhinorrhea, or refractory headache even when papilledema is absent. Early lumbar puncture for CSF opening pressure measurement is crucial to avoid unnecessary investigation. Weight reduction and medical therapy remain effective for most cases.

Conflicts of Interest

The author declare no conflicts of interest.

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