IDIOPATHIC NORMAL PRESSURE HYDROCEPHALUS WITH AUTONOMIC DYSFUNCTION DESPITE ATYPICAL NEUROIMAGING FINDINGS

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Abstract

Idiopathic normal pressure hydrocephalus (iNPH) classically presents with gait disturbance, cognitive decline, and urinary incontinence. A subset of patients may exhibit autonomic dysfunction, complicating diagnosis.

We present a 54-year-old male with progressive neurological decline and dysautonomia without ventriculomegaly, leading to delayed diagnosis.

This case highlights the need for clinical suspicion of iNPH despite atypical imaging and emphasizes the role of multidisciplinary evaluation.

Introduction/background

iNPH = gait disturbance + cognitive impairment + urinary incontinence
Diagnosis:

- Clinical + imaging (typically ventriculomegaly)
- Response to CSF diversion

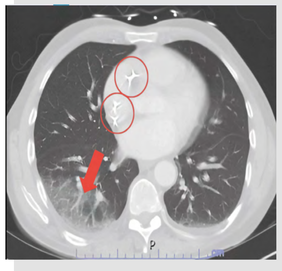
Dysautonomia occurs in ~1.5–3.7%

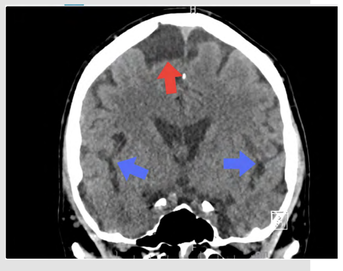
- Orthostatic hypotension
- Bradycardia
- Neurogenic bladder

Problem: Autonomic symptoms can mask iNPH → misdiagnosis

Case presentation

- 54-year-old male with history of psoriasis and hypothyroidism (on levothyroxine)
- 6–12 months of:
  i. Progressive gait instability
  ii. Cognitive decline
  iii. Urinary incontinence
- Presented with altered mental status
- Vitals: BP 90/60 mmHg, HR 45 bpm
- Labs and toxicology unremarkable
- Initially diagnosed with sick sinus syndrome with orthostatic instability
- Underwent dual chamber pacemaker placement and IV fluid resuscitation
- Temporary improvement → discharged
- Returned 2 weeks later with:
  i. Fever
  ii. Altered mental status
  iii. Gait instability
  iv. Urinary incontinence
  v. Bilateral lower extremity edema
- Workup:
  i. CMP and thyroid function tests normal
  ii. Blood cultures: 1/4 MSSA positive, repeat negative
  iii. Urinalysis: trace leukocyte esterase
- Treated as urosepsis with adrenal insufficiency
  i. Hydrocortisone and fludrocortisone
  ii. Antibiotics (cefazolin → nafcillin)
- Collateral history revealed:
  i. 1 year of personality changes and memory decline
  ii. Months of intermittent orthostasis and transient altered mental status
- Evaluation of pacemaker site showed swelling
- CT chest:
  i. Ground-glass opacity (right lower lobe)
  ii. Fluid collection near pacemaker → serom
- CT head:
  i. Frontoparietal parenchymal loss
  ii. Prominent Sylvian fissures
  iii. **No ventriculomegaly**
- Findings raised suspicion for atypical idiopathic normal pressure hydrocephalus (DESH variant)
- Neurosurgery consulted → ventriculoperitoneal shunt placed
- Post-procedure:
  i. Improvement in gait, urinary symptoms, and autonomic instability
  ii. Residual intermittent cognitive deficits





Discussion

iNPH pathophysiology involves:

- Impaired CSF dynamics
- Neurodegeneration
- Glymphatic dysfunction (AQP4 abnormalities)

Only ~50% of patients present with full Hakim's triad → diagnostic difficulty

Symptoms arise from:

- Ventricular expansion → parenchymal shear injury
- Compression of corticospinal and brainstem pathways

Subset of patients develop **autonomic dysfunction**:

- Orthostatic hypotension
- Bradycardia / sick sinus–like presentation
- Neurogenic bladder, constipation

Likely mechanism:

- Brainstem and diencephalic compression → autonomic dysregulation

Key challenge in this case:

- Dysautonomia dominated presentation → masked classic iNPH

Led to multiple misdiagnoses:

- Sick sinus syndrome
- Urosepsis
- Adrenal insufficiency

Atypical imaging:

- No ventriculomegaly
- Findings consistent with **DESH variant**

DESH can be misinterpreted as:

- Cortical atrophy → delayed diagnosis

Autonomic dysfunction likely contributed to:

- Orthostasis
- Edema (via vasodilation, reduced venous return)

Diagnosis supported by:

- Clinical pattern
- Imaging features
- **Response to VP shunt**

Post-shunt improvement confirms:

- Reversible component of disease

Conclusion

Maintain high suspicion for iNPH in patients with:

- Gait disturbance
- Cognitive decline
- Urinary symptoms
- **Prominent autonomic dysfunction**

Absence of ventriculomegaly **does not exclude iNPH**

Dysautonomia (orthostatic hypotension, bradycardia, urinary dysfunction) can:

- Mask underlying neurological disease
- Lead to diagnostic delays

Atypical presentations require:

- Thorough clinical evaluation
- Careful neuroimaging interpretation
- Broad differential diagnosis

Early recognition is critical:

- iNPH is **potentially reversible**

Multidisciplinary approach (neurology + neurosurgery) improves outcomes

Timely CSF diversion (VP shunt) → significant functional recovery

TAKE-HOME MESSAGE

- iNPH can present **without ventriculomegaly**
- Autonomic dysfunction may **mask the diagnosis**
- **Early recognition + VP shunt = reversible improvement**

Acknowledgements/references

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