Idiopathic normal-pressure hydrocephalus

Tina Hu MD MSc, Yuna Lee MD MEd

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1 Idiopathic normal-pressure hydrocephalus presents a diagnostic challenge

Although the complete triad is not seen in all cases, idiopathic normal-pressure hydrocephalus is characterized by gait dysfunction, cognitive impairment and urinary incontinence. These symptoms, however, are frequently encountered in older adults without the condition.[1] The onset of cognitive impairment in idiopathic normal-pressure hydrocephalus, which is often insidious with slow progression, may resemble Alzheimer disease,[2] but is potentially reversable.[3] The prevalence varies from 0.3% to 3% among patients older than 65 years, and increases with age.[3]

2 Gait dysfunction described as "freezing" and "magnetic" should trigger suspicion for the condition

Gait disturbance, typically the first symptom of idiopathic normal-pressure hydrocephalus, is characterized by a short-stepped gait and difficulty initiating movements, resulting in postural instability and falls.[1,3] Some gait abnormalities in the condition may mimic Parkinson disease, but idiopathic normal-pressure hydrocephalus is associated with a wide gait and normal arm swing.[3]

3 Magnetic resonance imaging of the brain showing ventriculomegaly with no obstruction to the flow of cerebrospinal fluid supports the diagnosis

Ventriculomegaly seen on imaging may be a result of progressive cortical atrophy. However, ventriculomegaly in the absence of, or out of proportion to, sulcal enlargement and cortical atrophy increases suspicion for idiopathic normal-pressure hydrocephalus.[1]

4 A high-volume lumbar puncture can be used as a confirmatory test and to select patients for surgery

The opening pressure is normal (8–18 mm Hg) in idiopathic normal-pressure hydrocephalus. Cognitive testing and a timed gait assessment should be conducted before and 30–60 minutes after the procedure.[2] Improved gait after a high-volume lumbar puncture (removal of 30–60 mL of cerebrospinal fluid) is a positive predictor of improvement with a ventriculoperitoneal shunt.[2]

5 A prompt referral to a neurosurgeon is recommended in selected patients

Ventriculoperitoneal shunts can improve symptoms, especially gait dysfunction, in up to 74% of patients.[4,5] Early treatment within 3 months of diagnosis is associated with better outcomes.[5]

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Affiliations: Department of Family and Community Medicine (Hu) and Division of General Internal Medicine (Lee), St. Michael's Hospital, Toronto, Ont.

Correspondence to: Tina Hu, tina.hu@mail.utoronto.ca