CLINICAL VIGNETTE

Normal Pressure Hydrocephalus

Susan Charette, MD and Patricia Harris, MD

Case Description

A 74-year-old man presented for an urgent office evaluation one day after a syncopal episode. The patient stated that he was in the kitchen and was trying to grab something from a cabinet above his head. He looked up, raised his arms and then started feeling dizzy. He experienced similar symptoms in the past and typically, they resolved spontaneously. This time he "blacked out" and woke up lying on his back on the floor.

The patient stated that his symptoms had mostly resolved, except any time he looked up with his head tilted back, he felt dizzy. He denied headache, confusion, muscle or joint pain, or lightheadedness. He reported no fever, chills, chest pain, or shortness of breath, no cough or urinary symptoms including urinary incontinence. His medical history includes coronary artery disease, atrial fibrillation, diabetes mellitus with peripheral neuropathy, hypertension, cervical degenerative disc disease status post C3-C6 discectomies and disc spacer placement, chronic lower back pain, and polycythemia vera. Over the last few months, he has complained of fatigue, trouble walking more than a block, and mild memory loss such as forgetting names.

His vital signs were normal including his orthostatic blood pressure measurement. His physical examination, including his neurologic assessment, was unremarkable except for his unsteady, wide based gait with a notable decreased step length**.** His EKG was normal and blood tests were unremarkable. Urgent computed tomography (CT) scans of the cervical spine and brain were obtained. CT scan of the cervical spine showed chronic findings related to his prior C3-C6 discectomies and disc spacer placement. The CT scan of the brain showed moderate normal pressure hydrocephalus.

The patient was referred to neurology for evaluation of his normal pressure hydrocephalus (NPH), his gait impairment, mild memory changes, and syncope. Based on his symptoms and the CT brain results, the neurologist scheduled inpatient large vessel lumbar puncture (LVLP), also known as the tap test. The tap test consists of a lumbar puncture followed by the removal of a large volume (typically 40–50 mL) of CSF and followed monitoring to see if the patient has an improvement in symptoms.[1] After this patient's LVLP, his walking returned to normal, and he decided to proceed with the CSF shunt placement. Normal pressure hydrocephalus is rarely a source of syncope and due to his multiple comorbidities, the patient scheduled follow up with his cardiologist and spine neurologist.

Discussion

Normal pressure hydrocephalus (NPH) is the classic trio of gait difficulty, cognitive impairment, and urinary incontinence. Neurosurgeon Dr. Salomon Hakim first published cases on hydrocephalus with normal pressure in 1964 and created the name "normal pressure hydrocephalus" based on his initial research findings.[2] The diagnosis of NPH is based on the patient's clinical history, physical examination, and brain imaging.[3] There are two types -- idiopathic NPH and secondary NPH. Idiopathic, or primary, NPH occurs without a clear cause. Secondary NPH is due to an underlying cause such as meningitis, head trauma, subarachnoid hemorrhage, central nervous system tumor and prior brain surgery.[3,4] Idiopathic NPH is more common and the focus of this article.

Men and women are equally affected by NPH.[1,5] The prevalence rate varies from 0.3% to 3% in patients greater than 65 years old.[3,4] Only 60% of patients with NPH present with the triad.[3] The signs and symptoms associated with NPH are common in other medical conditions. Gait impairment, cognitive changes, and urinary incontinence occur in 20% or more of men and women over 75 years old.[5]

Classically patients with NPH start with gait impairment along with either cognitive impairment or urinary dysfunction.[6] When patients present with only one or two of the NPH symptoms, it is important to ask if they are experiencing any others. The details of their symptoms should be pursued including their duration, severity, progression, prior diagnoses, and previous treatment. A review of the patient's medical history may reveal medical conditions with similar symptoms such as cerebrovascular disease, neurocognitive disorders (Alzheimer's, Parkinson's and Lewy body disease), spinal stenosis, peripheral neuropathy, vestibular problems and primary urological disorders.[1]

The physical examination is an important part of the NPH evaluation. Common gait findings in patients with NPH include widened stance, broad-based gait, reduced stride length, shuffling, impaired tandem, and bradykinesia or hypokinesia.[3,6] The gait may also have a reduced postural response similar to Parkinson's Disease, with a more frontal gait dysfunction including disequilibrium, abnormal stance, and postural adjustments.[3] For gait assessment, the simple Timed Up and Go Test or the more detailed *Performance Oriented Mobility Assessment (POMA*), also referred to as the Tinetti Test (*TT),* are recommended.[3] The impact on cognition is typically frontal

lobe executive dysfunction and psychomotor slowing.[3] These signs of dementia may include personality changes, loss of motivation, apathy, poor judgement, lack of impulse control, emotional outbursts, and depressed mood.[3] The Mini-Cog is useful for rapid cognitive impairment screening and the Montreal Cognitive Assessment (MoCA) is often used for more detailed cognitive screening. Urinary incontinence in patients with NPH usually results from detrusor muscle over-activity with symptoms of urgency, frequency and loss of urine.[3,6] When NPH is not diagnosed and treated early, these symptoms can progress, worsen and become difficult to resolve.[6]

Brain imaging, with either a computed tomography (CT) or magnetic resonance imaging (MRI) of the brain, is important for NPH evaluation.[3] For the diagnosis, imaging must show ventricular enlargement without signs of cerebrospinal fluid (CSF) obstruction or significant sulcal enlargement.[3] Ventricular enlargement may also occur with other dementias and is part of normal aging.[3] The Evan's index measures the ratio of the maximum width of the frontal horns of the lateral ventricles and the maximum internal diameter of the skull.[3] The Evan's index of 0.3 or greater confirms ventricular enlargement and supports the diagnosis of NPH.[1,4] At this time, there is no established role in the use of biomarkers for the diagnosis of NPH.[3]

The "gold standard" diagnostic test for NPH is the positive large volume lumbar puncture (LVLP).[1] A trial with a LVLP – the tap test – should be performed to confirm the diagnosis of NPH and determine if the patient will likely respond to the long-term treatment with placement of a ventriculoperitoneal (CSF) shunt.[5] Surgical CSF shunting with subsequent drainage is the first line therapy for NPH and is recommended only for patients who have a notable improvement in their gait and bladder control post LVLP.[3-5,7] The shunt is inserted with a proximal and distal catheter into the ventricular or lumbar subarachnoid space and the peritoneal cavity, respectively.[3] The success of CSF shunting ranges from 50% - 80 %, with positive effect on symptoms in the post-operative period.[1,3] Shunt complications include anesthesia and surgery complications, acute intracerebral hemorrhage, infection of the shunt, subdural hemorrhage, infection of shunt, subdural hematoma, subdural hygroma, seizure, shunt malformation, headache, hearing loss, tinnitus, oculomotor palsies, and damage to intra-abdominal organs.[4] If gait impairment was the first symptom, then there is a better chance for successful NPH treatment with ventriculoperitoneal shunting.[4] Patients with progressive NPH with advanced symptoms tend to be less responsive to treatment compared to those with milder symptoms and with less than 2 years of symptoms of duration.[1,4]

Early and accurate diagnosis of NPH can be instrumental in achieving a successful intervention and avoiding potentially severe and irreversible outcomes.[1] Shunt placement may have a high impact on a patient's quality of life. The timely treatment of NPH reduces frequent falls and associated head injury, subdural hematomas, trauma and hips fractures as well as the potential reduction of gait abnormality, cognitive impairment, and frequent falls incontinence.[5]

REFERENCES

1. **Relkin N, Marmarou A, Klinge P, Bergsneider M, Black PM**. Diagnosing idiopathic normal-pressure hydrocephalus. *Neurosurgery*. 2005 Sep;57(3 Suppl):S4-16; discussion ii-v. doi: 10.1227/01.neu.0000168185.29659.c5. PMID: 16160425.
2. **Wilson RK, Williams MA**. Normal pressure hydrocephalus. *Clin Geriatr Med*. 2006 Nov;22(4):935-51, viii. doi: 10.1016/j.cger.2006.06.010. PMID: 17000344.
3. **Nassar BR, Lippa CF**. Idiopathic Normal Pressure Hydrocephalus: A Review for General Practitioners. *Gerontol Geriatr Med*. 2016 Apr 20;2:2333721416643702. doi: 10.1177/2333721416643702. PMID: 28138494; PMCID: PMC5119812.
4. **Graff-Radford NR**. Normal pressure hydrocephalus. *Neurol Clin*. 2007 Aug;25(3):809-32, vii-viii. doi: 10.1016/j.ncl.2007.03.004. PMID: 17659191.
5. **Oliveira LM, Nitrini R, Román GC**. Normal-pressure hydrocephalus: A critical review. *Dement Neuropsychol*. 2019 Apr-Jun;13(2):133-143. doi: 10.1590/1980-57642018dn13-020001. Erratum in: *Dement Neuropsychol*. 2019 Jul-Sep;13(3):361. PMID: 31285787; PMCID: PMC6601311.
6. **Bluett B, Ash E, Farheen A, Fasano A, Krauss JK, Maranzano A, Passaretti M, Tang-Wai DF, Van Gerpen J, Alonso-Canovas A, Youn J, Malm J, Martino D; International Parkinson and Movement Disorder Society Normal Pressure Hydrocephalus Study Group**. Clinical Features of Idiopathic Normal Pressure Hydrocephalus: Critical Review of Objective Findings. *Mov Disord Clin Pract*. 2022 Nov 23;10(1):9-16. doi: 10.1002/mdc3.13608. PMID: 36704075; PMCID: PMC9847299.
7. **AbdelRazek MA, Venna N**. Ventriculoperitoneal-Shunt Placement for Normal-Pressure Hydrocephalus. *N Engl J Med*. 2017 Dec 28;377(26):e35. doi: 10.1056/NEJMicm1701226. PMID: 29281574.