Idiopathic normal pressure hydrocephalus dementia

When evaluating a patient with suspected idiopathic normal pressure hydrocephalus (INPH) and dementia, it is important to consider other possible causes of dementia that may present with similar symptoms. The following are some differential diagnoses to consider:

Alzheimer's disease (AD): AD is the most common cause of dementia. It is characterized by progressive memory loss, cognitive decline, and behavioral changes. Distinguishing AD from INPH can be challenging, as both conditions can cause cognitive impairment. Neuroimaging, such as MRI or PET scans, and biomarker testing (e.g., amyloid and tau proteins) may help differentiate between the two.

Vascular dementia: Vascular dementia occurs due to impaired blood flow to the brain, resulting in cognitive decline. It may present with gait disturbances, executive dysfunction, and a history of strokes or cerebrovascular disease. Vascular changes seen on neuroimaging, such as infarcts or white matter lesions, can aid in the diagnosis.

Parkinson's disease dementia: Parkinson's disease (PD) is a movement disorder that can also lead to dementia. Patients with PD dementia may experience gait disturbances, cognitive decline, and Parkinsonian motor symptoms (e.g., tremors, rigidity, bradykinesia). Parkinsonism features can help differentiate PD dementia from INPH.

Lewy body dementia: Lewy body dementia (LBD) is characterized by the presence of abnormal protein deposits called Lewy bodies in the brain. It can cause cognitive decline, visual hallucinations, fluctuations in attention and alertness, and Parkinsonian motor symptoms. Distinguishing LBD from INPH may rely on clinical features, such as prominent hallucinations and rapid eye movement (REM) sleep behavior disorder.

Frontotemporal dementia: Frontotemporal dementia (FTD) encompasses a group of disorders characterized by progressive changes in behavior, personality, and language abilities. Memory loss may not be as prominent in the early stages. Differentiating FTD from INPH involves careful assessment of behavioral and language symptoms, as well as neuroimaging findings.

Normal aging: Age-related cognitive changes can be mistaken for dementia. However, in normal aging, cognitive decline is mild and does not significantly impact daily functioning. It is essential to differentiate between normal aging and pathological causes of dementia

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At the time of the Idiopathic normal pressure hydrocephalus diagnosis, cognitive performances differed from cognitively healthy people in all CERAD-NB subtests. When the iNPH and AD patients' results were compared, the iNPH patients performed worse in Verbal Fluency and Clock Drawing tests while the AD group had more pronounced episodic memory dysfunctions. Nerg et al. demonstrated significant differences in the CERAD-NB subtests between cognitive profiles of iNPH and AD patients. These differences are not captured by the MMSE alone [1)].

Frontal and subcortical deficits (psychomotor slowing and impaired attention, executive, and

visuospatial dysfunction) can be the earliest cognitive signs of iNPH.

Significant improvement in these disturbances can occur after shunting.

More global cognitive deficits can be identified in individuals with suspected iNPH, even in those with Mini-Mental Status Examination (MMSE) scores greater than 25, and the severity of cognitive deficits appears to correlate with the presence of vascular risk factors.

Cerebrovascular disease is comorbid in over 60% of patients with iNPH.

Asymmetric resting tremor, lead pipe rigidity, or visual hallucinations may suggest dementia with Lewy bodies (DLB), which causes similar cognitive deficits. Depression with pseudodementia is in the differential diagnosis as well. Early presence of cortical deficits such as aphasia, apraxia, or agnosia should raise suspicion for dementia with cortical pathology, such as Alzheimer's disease (AD), multi-infarct dementia, or frontotemporal dementia. In patients with progressive dementia who lack gait dysfunction, a cause other than iNPH should be considered, regardless of ventriculomegaly.

Comorbid AD and iNPH is not uncommon, and the likelihood of each is increased with the presence of hypertension and advancing age. AD pathology is present in cortical biopsy of 75% of those iNPH patients with significant dementia at the time of shunt surgery.

Although gait can improve with shunting in such patients, dementia typically does not. Surgical treatment is generally discouraged for patients with severe dementia, even in the setting of gait dysfunction and incontinence, regardless of radiographic findings [2].

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Nerg O, Junkkari A, Hallikainen I, Rauramaa T, Luikku A, Hiltunen M, Jääskeläinen JE, Leinonen V, Hänninen T, Koivisto A. The CERAD Neuropsychological Battery in Patients with Idiopathic Normal Pressure Hydrocephalus Compared with Normal Population and Patients with Mild Alzheimer's Disease. J Alzheimers Dis. 2021 Apr 21. doi: 10.3233/JAD-201363. Epub ahead of print. PMID: 33896842.
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https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2674287/