Normal-Pressure Hydrocephalus: Past, Present and Future Management and the Relevance of New Insight of Neuropsychology Assessment for Measure of the Cognitive-Behavioral Sequelae

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Abstract

Normal Pressure Hydrocephalus (NPH) is a neurological disorder characterized by normal or mildly elevated intracranial pressure. Distinguishing NPH from other neurodegenerative disorders such as Alzheimer's disease and vascular dementia is challenging. Therefore, it may be undiagnosed and inappropriately managed. Patients with NPH are treated surgically with placement of a ventriculoperitoneal shunt; subsequently, most of them have shown clinical improvement in gait disturbance, bladder dysfunction, and cognitive functions. The case study within the present report aims to contribute new insights relative to the management and assessment of NPH. The present case additionally helps to inform the relevance of neuropsychological evaluation specific to the cognitive and behavioral sequelae of NPH. The case discussion provides data relative to the intersection of medical and psychological domains that further advance the classification and differentiation of NPH from other medical diseases or cognitive impairments. Further, the implementation of prospective neuropsychological assessments is discussed to help distinguish and inform more succinct diagnoses and appropriate treatment modalities for future providers managing the care of those suffering from NPH or NPH-like symptoms.

Keywords: Normal Pressure Hydrocephalus; Cerebrospinal Fluid; Lumbar Puncture; Idiopathic; Ventriculoperitoneal Shunt

Introduction

NPH is a neuropsychiatric disorder characterized by abnormal cerebrospinal fluid (CSF) build-up in the ventricles with no significant changes in the intracranial pressure (ICP). The disease was first described by Hakim and Adams in 1965 [1]. NPH is a potentially reversible syndrome characterized by enlarged ventricles, cognitive impairment, gait ataxia, and enuresis [2]. The insertion of a ventriculoperitoneal shunt is the current standard of care [3]. Its prevalence is estimated to be around 1.3% in people over 65 years old and increases with age [4]. NPH remains severely underdiagnosed; therefore, it is believed to be more prevalent than current data suggests amongst the elderly.

Various factors have been implicated in the pathogenesis, including abnormal CSF dynamics, abnormalities in both arterial and venous cerebral systems, neurodegeneration, inflammation, cerebral hypometabolism, and hereditary factors that influence CSF dynamics. Specifically, as detected by neuroimaging (i.e. MRIs), blockages in the absorption of CSF are among the neurophysiological issues noted amongst patients with NPH [5]. Nonetheless, it remains unclear whether CSF disturbances initiate and promote neural degeneration, but studies show that despite the presence of markers of neurodegeneration, NPH has a different CSF biomarker profile from Alzheimer's disease [5].

As CSF continues to accumulate the ventricles start dilating and compressing surrounding structures, the elevated pressure aggravates the cerebral hypoxia leading to disruption of normal metabolic processes [6] parenchymal scarring and stiffness, ultimately decreasing the brain’s compliance which is sometimes manifested with a mild increase of the intracranial pressure (ICP) [7].

Case Report

The present report is a case of a 68-year-old male who presented with a five-month history of pain in the lower extremities, gait ataxia. He underwent bladder surgery one year prior to his clinical presentation. The patient’s past history was significant for hyperlipidemia, osteoarthritis, and thyroid dysfunction. The patient has been seen by a podiatrist who concluded that the patient had plantar fasciitis, and prescribed steroid injections, to which the patient was unresponsive. Additionally, the patient tried physical therapy, which didn’t help either. Later on, the patient started to complain of lack of concentration, short-term memory loss and impaired attention.

Imaging

Physical examination

On examination, the patient’s lower back showed diffuse tenderness and diffuse moderate spasms. The Straight Leg testing was abnormal at 75 degrees/90 degrees. The manual muscle power testing was normal for all four extremities except the left ankle which was 4+/5, reflexes were symmetrical, gait was slow and ataxic and sensation was diminished to pinprick and touch on the left calf and left dorsal foot.

EMG examination

EMG was performed and revealed electrical evidence of bilateral L4 through S1 radiculopathy but no peripheral neuropathy.

MRI lumbar spine and brain

MRI of lumbar spine showed multiple disc herniation’s at T11-L1, L3-L4, L5-S1 and disc bulges from L1 to L3, L4 to S1. MRI of the brain revealed moderate enlargement of the ventricular system slightly out of proportion to mild-to-moderate prominence of the Sylvian fissures and sulcus patterns. Additionally, chronic ischemic changes were noted.

Mental status examination

Upon examination, the patient endorsed cognitive difficulties, including difficulty concentrating, distractibility, and short-term memory loss. The patient additionally endorsed psycho-emotional concerns, such as low energy, fatigue, anhedonia, worry, and irritability. He additionally appeared apathetic, as evidenced by his apparent impassivity and seemingly emotionless demeanor during the consultation.

Discussion

Evan's index, an indicator of ventricular volume, was above 30, which is considered to be in the pathological range and subsequently suggestive of ventriculomegaly [8]. There was no evidence of brain lesions on MRI. Lumbar puncture performed in this patient, revealed normal CSF pressure.

Additionally, a CFS tap test was performed and 40 - 50 ml CSF was removed. Evidence of improvement of symptoms in a couple of days. Concerning diagnostic impressions and rule outs, given the trajectory of symptom and disease progression, we rule out Alzheimer's Disease. Neuroimaging results did not reveal the canonical markers (i.e. neurofibrillary tangles, Tau deposits, atrophy of cortical regions) as would be expected from an individual suffering from AD. The patient's memory loss, which was chronic and progressive, has been associated with the NPH triad (i.e. cognitive issues, gait disturbance, incontinence).

Ventriculomegaly is a common finding in elderly patients. Therefore, NPH is not an MRI-based diagnosis and the only reliable diagnostic test for NPH is lumbar puncture with removal of 20 - 40 cc CSF. Improvement of patient's symptoms following CSF drainage favor the diagnosis of NPH. The findings of reduction in the brain parenchyma volume due to aging explain the enlargement of the ventricle as a response to this volumetric reduction. However, Alzheimer's can be associated with NPH and a diagnosis of exclusion must always be evaluated carefully.

Neuropsychological assessments

Variability in the etiology and the clinical presentation of NPH often compromise the ability to make diagnoses and, subsequently, impose the risk of delaying the discernment of the appropriate intervention. Therefore, the implementation of mental health provisions within the healthcare team may help alleviate such concerns. Mental health professionals, such as clinical neuropsychologists, can ascertain a patient's cognitive profile to assist in parceling out differential diagnoses and guiding the proper management. Neuropsychology is a sub-division of clinical psychology, wherein such providers examine neurocognitive status of the patients through the use of standardized assessment batteries.

Likewise, neuropsychological implementations offer a way of collecting data to test and monitor many of the cognitive domains (i.e. memory and attention domains of executive functioning) that are omnipresent within conditions such as NPH, as well as evaluate treatment efficacy with empirical quantitative data. No less, the inclusion of neuropsychological data can additionally be integrated to evaluate the effectiveness of treatment and monitor the patient's progress by collecting empirical quantitative data that is compared to normative samples for standardization. Moreover, given the diagnostic ambiguities that arise in presentations such as NPH, neuropsychological implementations (i.e. test scores on cognitive, memory, executive functioning, or psychomotor function), can offer an objective measure of functional impairments and diagnostic indices.

Neuropsychological assessments have demonstrated the capacity to track the progression of cognitive deficits. Evidence-based neuropsychological assessments, such as the Mini Mental State Examination (MMSE), offer the ability to not only rapidly and accurately screen for cognitive deficits, such as those present in dementia, but also favor specific diagnoses. Essentially, neuropsychology provides a fast, easy, reliable, non-invasive, and evidence-based modality to assist in diagnostics, treatment planning, and, ultimately, offers an objective measure of the cognitive-behavioral sequelae of NPH.

Common co-occurrences relative to cognitive decline amongst patients with NPH include detriments to the domains of memory, attention, information processing, psychomotor speed, visuospatial and visual constructional domains, and executive functioning [9]. As in the present case study, providers were otherwise unable to discern dementia from the natural course of aging concerning his memory deficits irrespective of medical diagnostic measures. However, cognitive domains specific to the various types of memory and memory-related processes can be easily identified using neuropsychological assessments.

It is not uncommon for patients with NPH to experience apathy and poor insight into their condition. The disease has been found to originate in subcortical regions of the frontal lobe of the brain, which disrupts neural circuitry responsible for psychomotor speed and emotional processing [9]. Hence, patients will experience "frontal-subcortical dementia" and present with mood symptoms, mental processing, and psychomotor speed delays.

Notwithstanding the patient's traumatic history of sustaining a gunshot wound incurred from an armed robbery assault compounded by his worsening symptomatology throughout his undiagnosed NPH, the case highlights the utility of integrative approaches

to providing care, encompassing mental health services coupled with neuropsychological data. Such implementations into patient care can help ensure timeliness and ease of diagnosis, as well as assist in identifying accurate and effective treatment interventions. Correspondingly, neuropsychological assessment may be necessary to ensure that patients recovering from NPH are realigned with premorbid levels of functioning. As demonstrated by the present case study, future healthcare providers may benefit from monitoring the cognitive and behavioral ramifications of NPH to ensure full recovery. Hence, it is of relevance to explore the residual psychological effects that may follow a diagnosis that imposes deficits and impairments in activities of daily living.

Conclusion

The psycho-emotional and cognitive implications are equally essential elements to the clinical picture with respect to implementing comprehensive and integrative diagnostic assessments and treatment planning. Imaging modalities such as CT and MRI reveal specific radiological signs that assist in establishing a final diagnosis. Furthermore, these modalities are increasingly being used to identify patients who would benefit the most from shunt implementation and to evaluate the surgical outcomes [10,11]. Considering the multifactorial etiology and as the underlying pathogenesis of NPH remains to be elucidated, the only current and definitive treatment is ventricular shunting. NPH is one of few conditions with dementia that can be successfully treated with as much as 80 to 90% of patients responding to shunt surgery leading to improvement in cognitive function and gait impairment [12]. Despite the fact that many aspects of this condition remain unexplained, NPH, for the most part, can be treated more successfully and has a better prognosis than other neurodegenerative diseases.

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