Idiopathic normal pressure hydrocephalus clinical features

Currently, there is no pathological hallmark for idiopathic normal pressure hydrocephalus [1)]

The natural course of iNPH is symptom progression over time, with worsening in gait, balance, and cognitive symptoms. This deterioration is only partially reversible.

Typically elderly presenting with gait abnormality, cognitive impairment, and urinary incontinence, with enlarged ventricles of the brain but normal or slightly elevated cerebrospinal fluid (CSF) pressure [2) 3)].

It can occur with varying combinations or degrees of each of the elements of the classic clinical triad first described by Hakim and Adams in 1965 [4)].

However, this complete triad is not always seen. Generally, gait disturbance plus one additional feature is required to consider the diagnosis

Postural stability in NPH is predominantly affected by deficient vestibular functions, which did not improve after the spinal tap test. Conditions which improved best were mainly independent of visual control and are based on proprioceptive functions [5)].

It is frequently present with cerebral vasculopathy; significantly increased prevalence of cardiovascular disease iNPH patients, which provides evidence that cardiovascular disease is involved as an exposure in the development of iNPH [6)].

Idiopathic normal pressure hydrocephalus (iNPH) may present, besides the classic triad of symptoms, extrapyramidal parkinsonian-like movement disorders.

Psychiatric manifestation of severe disabling anxiety [7)].

Abnormal sleep breathing is frequently associated with iNPH. Validation in larger series is required but Román et al. from the Houston Methodist Hospital and Weill Cornell Medical College, Cornell University, New York, suggest including sleep evaluation in patients suspected of iNPH [8)].

In SINPHONI—a Japanese multicenter cohort study looking at the validity of MRI findings in idiopathic NPH (iNPH) [9)] —there were only 51% of patients with the complete triad of symptoms. Sexual dysfunction [10)] neurological symptoms, psychiatric symptoms, or other infrequently reported signs have circumstantial relation to NPH but may hinder diagnostic processing [11)].

Gait disturbance

Idiopathic normal pressure hydrocephalus Gait disturbance

Urinary incontinence

Idiopathic normal pressure hydrocephalus urinary incontinence.

Dementia

Idiopathic normal pressure hydrocephalus dementia.

Apathy

Apathy is frequently observed in idiopathic normal pressure hydrocephalus (iNPH) and worsens cognitive impairment and gait disturbance. Chadani et al. evaluated the regions associated with apathy in iNPH using statistical imaging analysis on the whole brain, both in terms of cerebral blood flow and gray matter volume. Twenty-seven patients with iNPH were assigned to two groups based on their scores on the neuropsychiatric inventory items related to apathy; 18 patients were assigned to the group with apathy (iNPH + APA) and 9 to the group without apathy (iNPH - APA). The magnetic resonance images and cerebral blood flow single-photon emission computed tomography data of the two groups were compared using statistical parametric mapping 12. The regional gray matter volume of the right precuneus was significantly larger in the iNPH + APA group than in the iNPH - APA group, but the regional cerebral blood flow in any region of the brain was not significantly different between the two groups. These results suggested that the larger gray matter volume, which is thought to reflect gray matter compression, in the precuneus might be involved in apathy in iNPH [12)].

Comorbidities

Idiopathic normal pressure hydrocephalus Comorbidities

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