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STATE OF THE ART

Idiopathic Normal Pressure Hydrocephalus – What We Know

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ABSTRACT

Idiopathic normal pressure hydrocephalus is a neurological disorder with clinical diagnosis and imaging confirmation that benefits from a neurosurgical therapeutic solution in the vast majority of cases. Prevalence among the general population is not well defined at this time, but prevalence and incidence in developed countries is certainly steadily increasing due to increased life expectancy and evolution of diagnostic methods. Early diagnosis and appropriate treatment contribute to improving living conditions and can significantly reduce the cost of medical care for the elderly.

Keywords: normal pressure hydrocephalus, clinical diagnostic triad, dementia, neurosurgical treatment

INTRODUCTION

Idiopathic Normal Pressure Hydrocephalus (INPH) – first described as a diagnostic entity by Hakim and Adams in 1965, originally relied on the clinical triad of walking disorders – urinary disorders – cognitive impairment (1, 2). The development of new and exact imaging methods lead to the subsequent addition of CT and MRI diagnostic criteria, *i.e.*, dilation of the lateral ventricles, especially the frontal horns, with normal pressure in the CSF circulation spaces (3). Although the possibilities of imaging evaluation have developed and expanded in the last decades, the clinical diagnostic criteria remain the most important part of determining the diagnosis of INPH and selecting patients who could benefit from neurosurgical treatment (1, 2, 4).

Clinical Presentation

Walking disorders from INPH should be viewed through a wider prism as changes in both walking and posture. Walking characteristics in INPH patients include widening of the support base, dif-

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ficulty in starting the step, reducing of the step length and of the frequency of walking steps, difficulty in changing the direction of walking (1, 2).

Postural changes are embedded in the sphere of balance disorders with the difficulty or even impossibility of maintaining posture without a resting support.

Walking in NPH is considered as a walking apraxia, a combination of motility deficits: altered posture reflexes and fine adjustment of walking associated with ineffective inhibition of vestibular walking control center. This results in a "magnetic" run characteristic of INPH patients. There is a degree of hypertonia with live reflexes in the lower limbs, explained by the intimate proximity of cortical-spinal fibers with motor function for the lower limbs, at the corona radiata, with the external wall of the lateral ventricle. This explains the fact that the disturbing movements appear first in terms of chronology and also improve the first and in significant degree after surgical treatment (1, 2, 5).

Urinary disorders seen in patients diagnosed with INPH are incontinence, initially manifesting as micturition urgency, then gradually passing through different phases and culminating in complete urinary incontinence.

In advanced stages of the disease, anal sphincter incontinence can also be associated. If initially the urinary disturbances occur due to the involvement of sacral fibers of the cortical-spinal tract, also located in the radius of the corona, adjacent to the side-wall of the lateral ventricles, they later become an expression of the evolution of dementia that first overlaps and then follows the other manifestations of INPH (1, 2, 5).

If the first two manifestations in the symptomatic triad of Hakim-Adams have the same expression in most patients, although in varying degrees, the symptom that complements the clinical triad, namely cognitive disorders, can be difficult to fit into a single picture. Memory disturbances are similar to those found in other forms of dementia, especially major alterations of short-term memory. Slow ideation, speech difficulties (difficulty in finding words and pronouncing them), loss of interest in relating with the surrounding people and the environment. These changes in the ideatic, cognitive and relational status complement the clinical picture of the disorders encountered in patients with INPH (1, 2, 5, 6).

Imaging findings

The addition of imaging investigations – CT and mostly MRI – to clinical elements has helped to facilitate diagnosis and improve the sensitivity of the INPH diagnosis. The main changes of the CT and MRI examinations observed in these patients are the widening of the arachnoid spaces, the increase of the lateral ventricular volume, especially the frontal horns, and the increase of the cerebrospinal fluid (CSF) flow at the Sylvius aqueduct. These are the most frequent imagistic modifications in patients with INPH (3, 7).

The MRI exam performed as a routine investigation, with conventional spin echo and fast spin echo sequences, can be considered an im-

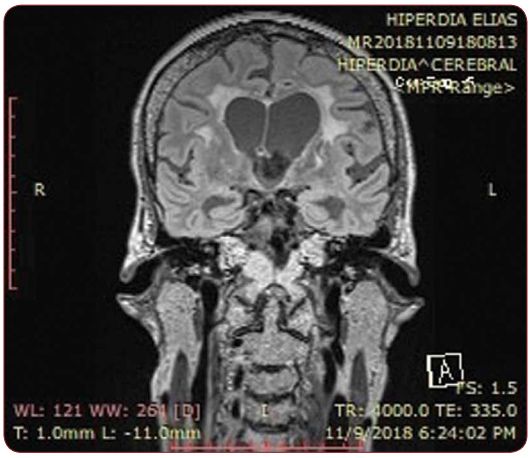

FIGURE 1. Coronal FLAIR MR shows enlarged lateral and third ventricles with interstitial edema along the lateral ventricles' margins

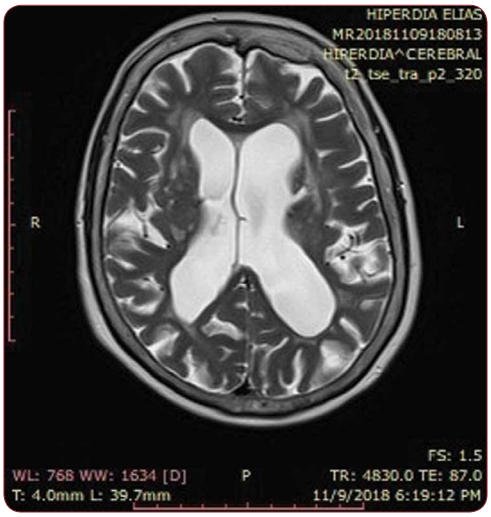

FIGURE 2. Axial T2 MR shows enlarged frontal horns of the dilated lateral ventricles with sulcal space dilation

portant factor for predicting the effectiveness of surgical treatment. Thus, the increase in CSF flow in the Sylvius aqueduct is a positive argument for neurosurgical treatment (3, 7, 8).

There are studies conducted through MR elastography over the last period that highlight a significant increase in cerebral rigidity in patients with INPH, but these investigations cannot guide a positive diagnostic or establish a therapeutic response yet (8, 9).

Pathophysiology

The advances in NPH imaging diagnosis have not unfortunately been coupled with advances in explaining the etiology of this diagnostic entity and, consequently, neither in the anticipation and prevention of NPH, as the vast majority of imaging modifications described above as found in patients with iNPH are frequently encountered in the routine examination of the elderly as well.

Over the last five decades several theories of episodic NPH have been advanced, none of which have been able to achieve unanimous acceptance.

The most common etiological theories of the INPH symptom complex were focused on the pressure and/or ischemic changes noted in these patients. On the one hand, the initial CSF circulation disorder at the level of arachnoid granulations (Pachioni) leads to a decrease in CSF reabsorption and therefore CSF accumulation in the ventricular system and subarachnoid spaces with their subsequent expansion, especially in compartments where the external resistance is low (mostly at the front horns). On the other hand, systolic/diastolic pressure variations in the ventricular system facilitate: 1) the steady-state transmission of the systolic wave on the ventricular walls that pose a lower resistance and 2) the repetitive whipping of the white matter fibers in their vicinity. In the view of some authors, these two factors would explain the etio-pathogenesis of INPH (5, 10).

With regard to the circulatory changes produced in these patients, which could be the cause of INPH, some are considered as possible triggers and maintenance and worsening factors. The alteration of the arteriolar wall structure is one of them (terminal-type circulation at the level of the cerebral white matter), with the significant reduction in the energy intake required for the proper functioning of parenchyma and cerebral cortex. The alteration of the venous return microcirculation is the other, with the disruption of the "wash" process at this level and the progressive accumulation of unnecessary degradation products that become cytotoxic by quantitative summation (1, 10).

Treatment

After establishing the INPH diagnosis, the next step is to devise a therapeutic attitude.

Surgical treatment, once the diagnosis of INPH established, consists of making a derivative of the circulatory CSF from the ventricular level, the unanimously accepted pathway nowadays being the peritoneal cavity (4, 11, 12). As the main step taken before surgery, a lumbar puncture is performed, thus releasing a quantity of CSF that varies depending on the experience and objectives set. This is usually around 30-50 mL, with concomitant CSF pressure measurement confirming normal pressure in the circulatory circuit of the CSF (13). The lumbar puncture may have a prognostic purpose, an important improvement of lumbar symptomatology being a positive prognostic factor for neurosurgical treatment (14). It also can have a diagnostic confirmation goal as the initial way of

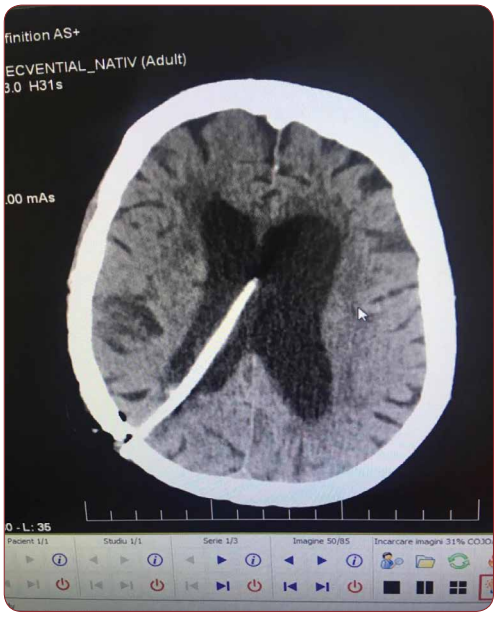

FIGURE 3. Axial CT scan shows correct placement of the ventricular catheter after surgical implantation and shunt

establishing this diagnosis, keeping in mind that S. Hakim treated the first patient diagnosed with INPH in 1965 with a lumbar puncture that relieved the symptoms and a subsequent ventricular-atrial derivation (1).

Neurosurgical treatment therefore consists of making a derivative of CSF circulation, the most accepted method being a ventricular-peritoneal shunt made with a reduced-flow (low pressure) anti-siphon valve. The manoeuver is performed under general anesthesia with oro-tracheal intubation, being a routine neurosurgical intervention (1, 2, 4, 11, 14).

The results of surgical treatment are evaluated after one month post-operatively by CT examination and clinical evaluation and then by MRI and clinical re-evaluation at SIX months and one year (1, 2, 4, 14). ❑

CONCLUSIONS

Although it is a relatively recent clinical and imaging entity, INPH is the only affliction from the wider framework of dementia benefiting from neurosurgical treatment. Studying causes, improving diagnostic methods and establishing new correlations between functional changes in the central nervous system, with additional attention to altering the production, circulation, and reabsorption of CSF processes could lead to significant relief of the suffering caused by these degenerative diseases that undergo a continuous increase in terms of incidence and social impact. ❑

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