Clinical Significance of Cerebrospinal Fluid Tap Test and Magnetic Resonance Imaging/Computed Tomography Findings of Tight High Convexity in Patients With Possible Idiopathic Normal Pressure Hydrocephalus

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Abstract

Idiopathic normal pressure hydrocephalus (iNPH) is a treatable syndrome with a classical triad of symptoms. The Japanese iNPH guidelines indicate that the cerebrospinal fluid (CSF) tap test and tight high-convexity on magnetic resonance (MR) imaging are important for the diagnosis. The relationships between the effectiveness of CSF shunt surgery in possible iNPH patients, the tap test result, and the MR imaging/computed tomography (CT) findings of tight high-convexity were evaluated in 88 possible iNPH patients (mean age 75 years) with one or more of the classical triad of symptoms, and mild to moderate ventricular dilation. All patients underwent the tap test in the outpatient clinic, and patients and caregivers assessed the clinical changes during one week. The tap test was positive in 47 patients and negative in 41 patients. Surgery was performed in 19 patients with positive tap test, and was effective in 17 patients. Although the findings were inconsistent in some patients, the result of the tap test was found to be highly correlated with the MR imaging/CT finding of tight high-convexity (p < 0.0001), confirming that both these diagnostic tests are promising predictors of shunt effectiveness.

Key words: normal pressure hydrocephalus, cerebrospinal fluid tap test, tight high-convexity, communicating hydrocephalus

Introduction

Normal pressure hydrocephalus (NPH) manifests as a syndrome consisting of one or more of the clinical triad of symptoms for NPH (gait disturbance, dementia, and urinary incontinence) and ventriculomegaly on magnetic resonance (MR) imaging or computed tomography (CT), associated with normal cerebrospinal fluid (CSF) pressure. CSF shunt surgery can reverse the symptoms dramatically.1,4) NPH can be classified into the idiopathic and secondary types. Idiopathic NPH (iNPH) is difficult to identify because of non-specific symptoms in elderly patients and the absence of preceding disorders.2) The Japanese Society of Normal Pressure Hydro-

cephalus published guidelines for iNPH in 2004,5,6) and included a flowchart for the diagnosis of the condition, based on evidence-based medicine, which requires amendment in the light of new evidence. The guidelines noted the diagnostic importance of the CSF tap test, but the incidence of patients with a positive tap test amongst suspected iNPH patients is not known. The guidelines also noted the diagnostic importance of high-convexity tightness on coronal MR imaging, but the correlation between the result of the CSF tap test and this specific finding on MR imaging/CT remains unclear.

The present study investigated the relationships between the effectiveness of CSF shunt surgery in
possible iNPH patients, the tap test result, and the MR imaging/CT findings of tight high-convexity.

Materials and Methods

This retrospective study involved 88 consecutive patients, 52 males and 36 females with mean age of 75.9 ± 5.47 years for males and 75.2 ± 5.99 years for females, with no statistical difference, admitted with possible iNPH over a period of 38 months between May 2004 and June 2007. The patients had one or more of the clinical triad (gait disturbance, dementia, and urinary disturbance), mild to moderate ventriculomegaly, and normal CSF pressure. The diagnosis of possible iNPH was established according to the Japanese guidelines for iNPH. Spinal tap was performed in all 88 patients at the outpatient clinic in the Department of Neurosurgery, Kitano Hospital. Patients who were taking part in a multicenter cooperative study were not included.

After measuring the CSF pressure via the spinal tap, 30 ml of CSF was drained spontaneously. The patients and their caregivers were asked to assess any changes in clinical symptoms that occurred during the following week. Transient or long-lasting improvements of any of the symptoms of the classical triad were regarded as a ‘positive tap test.’ Most improvements were related to gait disturbance. Quantitative assessments such as the timed up-and-go test or the Mini-Mental State Examination were not included in this outpatient clinic-based study. Four patients with marked ventriculomegaly indicative of long-standing overt ventriculomegaly in adults were excluded from the study.

Narrowing of the subarachnoid space in the high-convexity region (high-convexity tightness) was easily observed on coronal MR imaging (Fig. 1). Tight high-convexity was concomitant with tightness in the medial subarachnoid space, especially in the posterior half, in iNPH patients, and the latter finding could be assessed on the highest axial section of MR imaging/CT. In the present study, narrowing in the high-convexity and medial subarachnoid spaces was described as ‘tight high-convexity.’

The MR imaging/CT findings were not quantitative, so were assessed by the senior author (M.I.) and the findings were divided into (+) or (−). CSF shunt surgery was recommended for possible iNPH patients with a positive tap test. Ventriculoperitoneal shunt was performed with the Codman-Hakim programmable valve (Codman, Johnson & Johnson, Tokyo). Postoperative follow up was continued for more than 3 months. Shunt effectiveness was defined as postoperative improvement of any of the symptom(s) of the classical triad. Statistical analysis used PRISM software (version 4 for Macintosh; GraphPad Software, Inc., La Jolla, Calif., U.S.A.).

Results

The tap test was positive in 47 patients and negative in 41 patients. Only male preponderance was significant in the tap test-positive group (positive group: 53.4% vs. negative group: 46.6%, p < 0.05). There were no statistical differences in age (75.9 ± 5.21 vs. 84.8 ± 6.79 years), volume of CSF removed (24.5 ± 1.12 vs. 26.3 ± 0.91 ml), CSF pressure (11.7 ± 3.76 vs. 11.7 ± 3.59 mmH2O), or Evans index (0.32 ± 0.83 vs. 0.32 ± 0.13). No statistical differences were noted in symptoms of gait disturbance (100% vs. 100%), dementia (55.3% vs. 51.2%), and urinary incontinence (33.3% vs. 31.7%). Ventriculoperitoneal shunting was performed in 19 of the 47 patients with positive tap test, and was effective in 17 of these 19 patients. Therefore, CSF shunt surgery was not performed in 28 of the patients with positive tap test, in most patients or their families did not wish for the procedure, but because of high surgical risk in two patients.

Tight high-convexity on MR imaging/CT was noted in 42 of the 47 patients with positive tap test, and in 16 of the 41 patients with negative tap test (Fig. 2). Chi-square analysis showed a statistically significant correlation between tap test result and tight high-convexity (p < 0.0001). However, some inconsistencies were also noted: negative tap test was found with tight high-convexity in 16 patients (Fig. 3), and positive tap test without tight high-convexity in 5
Fig. 2 Relationships between the results of the cerebrospinal fluid (CSF) tap test, findings of tight high-convexity, and outcome of ventriculoperitoneal (VP) shunting.

Fig. 3 Coronal T₂-weighted magnetic resonance images showing ventricular dilation and tight high-convexity. This 74-year-old male suffered from progressive short-stepped gait, but the tap test was negative. Continuous lumbar cerebrospinal fluid drainage also failed to result in any improvement in symptoms.

Fig. 4 Coronal T₂-weighted magnetic resonance images showing no tight high-convexity. This 72-year-old male suffered from recent development of short-stepped gait, which improved after the tap test. Ventriculoperitoneal shunting was effective.

Discussion

The present study investigated the effectiveness of CSF shunt surgery for possible iNPH, and the relationship between the result of the CSF tap test and the presence of tight high-convexity on MR imaging/CT. Forty-seven of the 88 patients with possible iNPH had positive tap test. There was no statistical difference in the initial CSF pressure and the volume of CSF removed between the positive and negative tap test groups. Nineteen of the 47 patients with positive tap test underwent CSF shunt surgery, and the procedure was effective in 17 of the 19. Therefore, a positive tap test is a good predictor of shunt effectiveness.

More than half of the patients with a positive tap test did not undergo shunt surgery. This surgical option was rejected by the patients themselves or their families. Almost all patients who experienced patients (Fig. 4).

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marked improvement after the tap test wanted to undergo surgery, supported by their families, and shunt effectiveness in these patients was remarkable. In contrast, the improvement was mild or short-lived for many of the patients who did not undergo surgery, so it might have influenced the expectations for CSF shunt surgery in these elderly patients or their families. Patients and their family were hesitant to accept surgery, but still wanted to achieve improvement. In these situations, marked improvement after the tap test prompted them to undergo surgery. Thus, the tap test is quite important both for the diagnosis, and for acceptance of the surgery. During the study period, no data were available to predict the surgical outcome for patients with mild or very short-term improvement after the tap test. Recently, assessment of the classical triad with the iNPH grading scale before and after the tap test was found to be valid for evaluation of the response on the tap test in in-patients. This scale would be useful for assessing the severity of symptoms semiquantitatively, even in the outpatient clinic. The diagnostic importance of narrowing of the subarachnoid space in the high-convexity and medial subarachnoid spaces in iNPH was investigated using MR imaging volumetry in association with mild to moderate ventriculomegaly. In the present study, such findings were described as ‘tight high-convexity.’ Tight high-convexity could be observed in both the coronal and highest axial sections of MR imaging/CT.

Tight high-convexity is important for the differentiation of iNPH from brain atrophy, but the relationship to the result of the CSF tap test was not known. The present study revealed a statistically significant correlation between the two, but also some inconsistency. If patients with tight high-convexity have a negative tap test, the differential diagnosis between iNPH and other senile neurodegenerative or vascular disorders, including progressive supranuclear palsy and corticobasal degeneration, becomes difficult. In contrast, the present study included 5 of 88 patients (5.7%) with positive tap test but without tight high-convexity. Shunt surgery was effective in all 3 of these patients who underwent surgery. Therefore, no finding of tight high-convexity on CT or MR imaging in iNPH patients is not well understood. Adhesion in the high-convexity region or upward displacement of the brain due to ventricular dilation are possible causes. After CSF shunt surgery, we usually observed mild improvement of tight high-convexity, and dilation of the ventricular system and the sylvian fissure tended to normalize. Arachnoid granulations in the high-convexity region are believed to be the site of CSF absorption. Tight high-convexity associated with dilated sylvian fissure and ventriculomegaly, which are typical findings of iNPH, may be due to impairment of CSF absorption by malfunctioning arachnoid granulations or adhesions in the high-convexity region.

iNPH can be regarded as a special form of communicating hydrocephalus, which we call disproportionately enlarged subarachnoid-space hydrocephalus (DESH). Recently, increased brain pulsation due to undamped arterial pulse pressure in the subarachnoid space has been described as important in the etiology of communicating hydrocephalus. The hydrodynamic theory is useful for understanding the pathophysiology of ventricular dilation in communicating hydrocephalus. However, whether the hydrodynamic theory explains the development of DESH is not known. Based on classical CSF bulk flow theory, DESH may be a typical form of communicating hydrocephalus due to impairment of CSF absorption at the arachnoid granulations in the high-convexity region. Further study is necessary to elucidate the

Other drawbacks of the tap test are the rare complications of bleeding, neural injury, and postural headache. Explanation and informed consent are necessary. Therefore, the tap test is useful, but not yet conclusive in some patients. Diagnosis based on MR imaging/CT is superior to that based on the tap test in terms of non-invasiveness, but the overall diagnostic superiority remains to be clarified.

The 3-day CSF external lumbar drainage is reported to have a higher diagnostic accuracy than the CSF tap test. This suggests that the continuous CSF drainage test might be more useful for decreasing false negative cases than the tap test. We performed this procedure for one patient who had progressive short-stepped gait with ventriculomegaly and tight high-convexity (Fig. 3). The tap test was negative and the external CSF drainage test for one day was also negative and accompanied by postural headache. Continuous external lumbar drainage appears to be more invasive for elderly patients with mild or moderate dementia, so is regarded as optional in the Japanese guidelines.5,6 The reasons for tight high-convexity on CT or MR imaging in iNPH patients is not well understood. When a patient has tight high-convexity, the differential diagnosis between iNPH and other senile neurodegenerative or vascular disorders, including progressive supranuclear palsy and corticobasal degeneration, becomes difficult. In contrast, the present study included 5 of 88 patients (5.7%) with positive tap test but without tight high-convexity. Shunt surgery was effective in all 3 of these patients who underwent surgery. Therefore, no finding of tight high-convexity on CT or MR imaging in iNPH patients is not well understood. Adhesion in the high-convexity region or upward displacement of the brain due to ventricular dilation are possible causes. After CSF shunt surgery, we usually observed mild improvement of tight high-convexity, and dilation of the ventricular system and the sylvian fissure tended to normalize. Arachnoid granulations in the high-convexity region are believed to be the site of CSF absorption. Tight high-convexity associated with dilated sylvian fissure and ventriculomegaly, which are typical findings of iNPH, may be due to impairment of CSF absorption by malfunctioning arachnoid granulations or adhesions in the high-convexity region.

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pathophysiology of iNPH.

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References


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Commentary

Ishikawa and collaborators analyze an interesting finding in adult normal pressure hydrocephalus (NPH). Tight high convexity (THC) of subarachnoid spaces of the brain as identified by MRI is shown to be a symptom associated with NPH. As of yet we do not know the cause of NPH and we can only improve its symptoms in some cases. This analysis is important, as it sheds light on morphological changes of the brain in NPH, which have so far not attracted much attention. The THC of subarachnoid spaces and the tap test were significantly correlated beyond any reasonable doubt, but there were also clear exceptions as a positive tap test and effective surgery were also observed in the absence of THC. Obviously our current concept of NPH comprises quite heterogeneous conditions and a promising idea is to look at better and more detailed imaging techniques to improve our understanding of NPH. As of now we are grateful to Ishikawa and his working group for this information on THC in NPH.

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Firstly, the authors further clarify the MR imaging/CT findings of patients with idiopathic normal pressure hydrocephalus. Secondly, they concluded that we can use both MR imaging/CT findings and the result of the tap test to predict the shunt effectiveness for these patients. These findings will give neurosurgeons a new tool to decide whether CSF shunt surgery should be done for patients with mild or very short-term improvement after the tap test.

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